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DISEASES OF THE SPINAL CORD



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DISEASES

OF THE

SPINAL CORD

BY

BYROM BRAMWELL, M.D., F.R.C.P. ED., F.R.S. ED.

ASSISTANT PHYSICIAN TO THE EDINBURGH ROYAL INFIRMARY AND LECTURER ON THE PRINCIPLES AND PRACTICE
OF MEDICINE IN THE SCHOOL OF MEDICINE, EDINBURGH; LATE PATHOLOGIST TO THE EDINBURGH
ROYAL INFIRMARY, PHYSICIAN AND PATHOLOGIST TO THE ROYAL INFIRMARY, NEWCASTLE-
ON-TYNE, AND LECTURER ON CLINICAL MEDICINE AND PATHOLOGY IN THE UNIVERSITY
OF DURHAM COLLEGE OF MEDICINE, NEWCASTLE-ON-TYNE; FORMERLY, MEDICAL
OFFICER TO THE TYNEMOUTH UNION WORKHOUSE HOSPITAL, THE PRUDHOE
MEMORIAL CONVALESCENT HOME, THE TYNE FLOATING HOSPITAL;
MEDICAL OFFICER OF HEALTH AND POLICE SURGEON TO
THE BURGH OF TYNEMOUTH; LECTURER ON MEDICAL
JURISPRUDENCE IN THE UNIVERSITY OF
DURHAM COLLEGE OF MEDICINE,
NEWCASTLE-ON-TYNE,
ETC., ETC.

WITH ONE HUNDRED AND SEVENTY ILLUSTRATIONS

THIRD EDITION

EDINBURGH
WILLIAM F. CLAY, 18 TEVIOT PLACE

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PREFACE TO THE THIRD EDITION

ALTHOUGH the Second Edition of my Diseases of the Spinal Cord has been out of print for several years, I have been so much occupied with other matters that I have not been able to re-issue the work until the present time.

In this Edition the subject-matter has been thoroughly revised and greatly extended; in fact, the text has been entirely rewritten and rearranged in the lecture form. A large number of new illustrations—chiefly clinical—have been added.

I trust that in its present form the work will meet with the same favourable reception which was accorded to the previous editions.

B. B.

23 DRUMSHEUGH GARDENS,
EDINBURGH, *April* 1895.

PREFACE TO THE SECOND EDITION

THE very favourable reception which the First Edition of this work received from the medical press both in this country and abroad, and the fact that it has been translated into the German, French, and Russian languages, have induced me to make fewer changes in this, the Second Edition, than I at one time contemplated.

But while the original plan of the work has been as far as possible adhered to, every page has been subjected to careful revision. The sections devoted to the pathology of individual lesions have been placed under the special diseases, the functional affections have been more fully considered, while a considerable space has been given to the important and difficult subject of concussion of the spine and the method of examining 'railway cases.' Many of the old illustrations have been redrawn, and a large number of new figures, both woodcuts and chromolithographs, has been added.

B. B.

23 DRUMSHEUGH GARDENS,
EDINBURGH, *October* 1884.

PREFACE TO THE FIRST EDITION

IN the following pages, which are based on a portion of my course of Lectures on Medicine, I have endeavoured to give a concise description of the more important points relating to the Diseases of the Spinal Cord.

In some places the original form is retained, but for the most part I have, for the sake of condensation, abandoned the colloquial style of the lecture-room, and in many places arranged the matter in the form of headings.

Believing that one great secret of all successful teaching is to teach by the eye as well as by the ear, I am in the habit of copiously illustrating my lectures by diagrams, drawings, and microscopical preparations. The diagrams and drawings are introduced into the text in the form of woodcuts, the microscopical sections are represented in colours.

The chromo-lithographs are all drawn by myself, first with the camera lucida, and then in lithograph chalk; they are with two exceptions (figures 56 and 151, which are copied from Charcot) representations of my own sections.

I am indebted to Professor Dreschfeld of Manchester, Dr. Robertson of Glasgow, Dr. Crease of South Shields, Dr. Banham of Sheffield, Dr. Goyder of Newcastle, and Dr. Milner Moore of Coventry, for spinal cords, from which some of the sections were made; and I am particularly indebted to Dr. D. J. Hamilton, not only for material, but also for much valuable instruction in the methods of investigating nervous structures.

My thanks are also due to Professor Charcot, Professor Flower, Professor Ferrier, Dr. Ross, Dr. Gowers, and Dr. Herbert Tibbits, for their kindness in allowing me to reproduce some of the figures which have appeared in their respective works.

A considerable portion of the first two chapters was published in the *Transactions* of the Northumberland and Durham Medical Society for November and December 1881.

B. B.

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LECTURES ON THE DISEASES OF THE NERVOUS SYSTEM

VOL. I.—SPINAL CORD

CORRIGENDA

Page 146, nine lines from the bottom—instead of ‘several other cases’ *read* ‘at least one other case.’

Fig. 43.—The nerve cells in this figure are rather too large.

Fig. 55.—Instead of ‘The deltoids are well preserved’ *read* ‘The deltoids are also atrophied.’

Page 306, line twenty-one from the bottom—instead of ‘usually firm and well nourished’ *read* ‘usually firm and well nourished in the early stage of the disease.’

Page 361, nine lines from the top—instead of ‘Thomson’s’ *read* ‘Thomsen’s.’

Page 522, six lines from the top—instead of ‘Several other cases of the same kind have’ *read* ‘At least one other case of the same kind has.’

The diseases of the nervous system are very numerous; many of them are rare; some of them are, as yet, very inadequately and imperfectly understood. In my opinion it is a mere waste of time to consider, in a course of systematic lectures, the rare and imperfectly differentiated forms of disease. What students and general practitioners want is a good working knowledge of those diseases which they are likely to meet with in ordinary everyday practice. The man who has this knowledge will be able to deal with the rarer forms of disease when he comes

¹ The diseases which were omitted from the course of lectures are treated of (in the lecture form) in the text.

across them, which, in the case of some diseases of the nervous system at all events, may be only once or twice in a lifetime.

My object, then, in this course of lectures will be to impart to you an *intelligent* knowledge of the more common and important diseases of the nervous system. Now, by an intelligent knowledge I do not mean a mere parrot knowledge. It would perhaps be possible in fifty lectures to *enumerate* the chief characteristics of every nervous disease; but at the end of such a course of lectures the student would have no intelligent knowledge of the subject; his mind would be simply stuffed with a confused mass of facts and details. What I want to give you is a knowledge which will enable you to appreciate and understand the manner in which the symptoms are produced, and the course which the diseased phenomena are likely to pursue; a knowledge which will enable you to distinguish the individual diseases from one another; and above all, a knowledge which will help you to intelligently deal with the diseases of the nervous system in the way of treatment.

If I succeed in imparting to you even a, comparatively speaking, limited amount of knowledge of this kind, I shall be content; for, as I have already said, a real, thorough and intelligent knowledge of the more common forms of nervous disease will enable you to deal with the rarer forms should you happen to come across them. If you get a grasp of the great principles of neurology and if you acquire an intelligent knowledge of the ordinary, typical forms of disease, you will have laid a solid basis of neurological knowledge.

Importance of Clinical Study.—And here let me impress upon you the importance of neglecting no opportunity of making yourselves practically familiar, in the living patient, with the diseases, which it will be my business, in this lecture-room, to describe to you, in a systematic manner. It is essential that your clinical and systematic studies should go hand in hand. It is impossible to obtain a real knowledge of disease from mere written descriptions, or from lectures, however interesting and lucid. The knowledge acquired by mere book-study may serve the purpose of an examination; but often (I will not say alas, for I do not regret it), as the mere book-student finds out to his cost, it is inadequate even for that purpose. Knowledge which is based on such a method of learning quickly disappears and is of little

or no use for the purposes of practice. Even those of us who are constantly dealing with nervous diseases in practice find it difficult to remember all the facts connected with the rarer forms of disease which we seldom meet with; and, for my own part, I do not profess to remember them. Some men have better memories than others, but there are very few memories which can stand such a strain as this. It is absurd, therefore, to expect students to remember these details; and, as I have more than once told you, it is, in my opinion, an abuse of examinations to examine men on rare diseases which they have never seen, and which they perhaps may not have an opportunity of seeing for years after they have entered practice.

Importance of Anatomy, Physiology, and Pathology.—The nervous system is the most complicated mechanism in the body; its anatomy and physiology are, as yet, only very imperfectly understood; this is one of the reasons why the study of nervous diseases is so difficult. The most satisfactory way of obtaining an intelligent knowledge of any disease—and the statement applies with special force to the diseases of the nervous system—is to endeavour to realise in the mind's eye:—*firstly*, the exact position and character of the lesion, which is the pathological substratum of the disease under consideration; and *secondly*, the effects (mechanical and vital or physiological) which that lesion is calculated and likely to produce in the part which is directly implicated, (the *direct* effects of the lesion), in the more distant parts of the nervous system, and in the other organs and tissues of the body (the *remote* effects of the lesion).

Anatomy, physiology, and pathology in its widest sense, are the only foundations on which an intelligent comprehension of the diseases of the nervous system can be based. I cannot, therefore, too forcibly impress upon you the necessity of making ourselves thoroughly familiar with these points in the anatomy, physiology and pathology of the nervous system, which are more directly important for the purposes of medicine. In addition, if you wish to obtain a firsthand knowledge of disease—and it is of course the only knowledge worth having—you must, as I have already told you, take every opportunity of observing the effects of disease in the living patient. Further, you should endeavour to follow out the results of disease in the dead-house;

you will not advance far in the study of neurology unless you try to follow up, in individual cases, the exact relationship between the symptoms in the living and the conditions which are found in the dead.

The True Method of Systematic Teaching and Study.—In describing the individual diseases of the nervous system, I shall direct your attention very specially to what I term the *pathological physiology*. Those of you who have attended my previous lectures know that, whenever it is possible to do so, I preface my description of the clinical phenomena with a brief account of the more important facts connected with the morbid anatomy and pathology of the disease; and that I make a special point of trying to explain the manner in which the different symptoms are produced, in other words, the relationship which exists between the lesion and the clinical phenomena which are the effects of that lesion. This method is, in my opinion, the basis of all intelligent teaching; for it is the only method which enables us to understand the whys and wherefores of disease. The worst kind of teaching consists in a bare enumeration of facts; such teaching is in fact no teaching at all. The only satisfactory method of studying the diseases of the nervous system is to endeavour to realise how the different clinical phenomena (the symptoms) are produced. If you make a point of trying to discover an explanation for the different clinical phenomena which come before you, if you refuse to allow yourselves to rest content with a bare knowledge of the facts, you will become better observers and you will easily and readily acquire a genuine knowledge of the subject that you have in hand. Such a method of learning is obviously much more interesting than the mere committing to memory of a mass of facts and details; but far more important than this, it is the only method which will enable you to acquire with ease and to remember with facility the complicated phenomena with which the neurologist has to deal. Unfortunately, this method of study cannot always be followed; for our knowledge is, as yet, imperfect, and there are many facts, gathered by the practical observation of disease at the bedside and in the dead-house, for which we have as yet no intelligent explanation. For example the facts that the sclerosis of the posterior columns of the spinal cord, which is the pathological substratum of locomotor ataxia,

usually tends to progress steadily from bad to worse, notwithstanding treatment; that iodide of potassium is a specific in tertiary syphilis, are facts which at present we are unable to explain but which are of vast importance. They are illustrations of a great mass of facts which you must learn and know. Isolated (unexplained) facts of this kind are difficult to remember, but it is essential that you should (by frequent repetition) make yourselves familiar with them.

Classification of Nervous Diseases.—We are in the habit of dividing the diseases of the nervous system into:—

(1) The diseases of the *brain*, including the cerebellum, the pons Varolii and medulla oblongata (although the pons Varolii and medulla oblongata are very different, both physiologically and pathologically, from the cerebrum or brain proper);

(2) The diseases of the *spinal cord*; and

(3) The diseases of the *peripheral* (cerebral, spinal and sympathetic) *nerves*.

This division, which is anatomical, cannot always be rigidly followed for the purposes of clinical medicine. Although the spinal cord and its peripheral nerves constitute a part of the nervous system which, for many physiological, pathological and clinical purposes, can be isolated from the brain; and although the brain can functionate independently of the spinal cord and spinal nerves, many of the brain functions can only be carried on with the aid of the lower portions of the nervous system (pons Varolii, medulla oblongata, spinal cord and peripheral nerves).

There is, I think, reason to suppose that almost every change in one part of the nervous system induces some sort of change in every other part; and that impressions, which enter the nervous system at the periphery, and functional changes which originate in the nerve centres without any *obvious* external exciting cause, are apt to reverberate, it may be in a very feeble manner, throughout the whole nervous system. Although the functional changes which are produced in this way may, for the most part, be too delicate to produce any obvious external effects by which they can be seen or recognised, it is important to realise that action and reaction are constantly going on throughout the whole nervous system, in health and also in disease. If this is so, you will easily understand that lesions

which can be recognised by the naked eye or the microscope (alterations which are very coarse and gross in comparison with those to which I have just referred) will in many cases produce marked disturbances in parts of the nervous system which are far removed from their primary seat.

My object in making these statements is to impress you with the idea that in studying the diseases of the nervous system it is essential not to concentrate the attention too exclusively upon the lesion and its immediate (direct) effects—though that is, of course, the first and most important point—but to regard the condition of the whole nervous system, and in fact the condition of the whole body. The nervous system is the great controller and regulator of organic function. Disease of the nervous system, especially the diseases of the higher nerve centres (of the brain) may produce disturbances in every organ and tissue of the body, for all parts of the body are represented or re-represented in the brain. Further, disease at the periphery may act and react upon the nerve centres (spinal cord and brain) and produce cerebral and spinal derangements and consequently cerebral and spinal symptoms. It is hardly necessary to give specific illustrations of these great truths. I may simply point out that vomiting is a common symptom in cases of cerebral disease; that mental depression is often due to disturbance of the function of the liver; that epileptiform convulsions are frequently due to uræmia; that glycosuria not unfrequently results from localised disease at the base of the brain; and that derangement of the function of the liver, heart, and probably of every organ of the body, may result from a mere psychical impression (such as profound grief, bad news, etc.,) which disturbs the higher cerebral (psychical) centres and then reacts upon the peripheral tissues and organs of the body.

Importance of the Personal Equation.—Another point which it is very important to remember in connection with the diseases of the nervous system is this, that individual patients differ very notably one from another. In former lectures I have repeatedly emphasised the fact that the various morbid processes which we term diseases, should not be regarded as mere morbid entities, but rather as derangements of function and structure occurring in *individual*, living, human beings. I cannot too frequently repeat that one and the same lesion or functional

disturbance may produce very different results (symptoms) in different individuals. In dealing with the diseases of the nervous system, the peculiarities of each individual patient have to be carefully studied. The personal equation, as it has been termed, is of special importance for the purposes of prognosis and treatment. I lay stress upon this point, for I presume that your object in attending this course of lectures is not merely to study the subject from a theoretical standpoint, but to acquire a working knowledge which will be helpful to you in practice.

Now, heredity is one of the factors in the personal equation which has to be taken into account. Heredity plays a very special and important part in the production of many diseases and functional derangements of the nervous system.

Further, you must remember that the more complicated the mechanism and the more highly differentiated and specialised the function of an organ, the more easily is its structure damaged and its function deranged and disturbed.

These statements apply with special force to cerebral diseases and mental derangements. The structure and function of every other organ of the body are simple in comparison with the structure and function of the brain. The physician can, in most cases, determine with considerable accuracy whether the liver, heart, or kidneys conform to the normal (average) standard of health or not. It is very different in the case of the brain. There is no normal (average) standard of mental activity and mental capacity; each individual is the standard for himself. These individual differences are often a source of difficulty in the diagnosis of mental derangements. Unless one is acquainted with the normal condition of brain function and development in a particular patient (his normal condition as regards intellectual capacity, memory, emotional control, moral feelings and the like), one may fail to detect the presence of some unmistakable mental derangement. Of course, such gross disturbances as acute mania or profound melancholia are obvious enough; it does not require a doctor to recognise those forms of madness. But the slighter forms of mental derangement, which may be quite as important, can, in many cases, only be recognised by comparing the mental condition of the individual, in his deranged or diseased, with his mental condition in his normal state.

I repeat that the personal equation and the hereditary tendencies of each individual patient are of the greatest importance in many diseases and derangements of the nervous system.

The Chief Forms of Organic Lesion.—The diseases and derangements of the nervous system are, for the most part, due to the same forms of lesion that occur in other tissues and organs of the body.

The function of the nerve centres may be disturbed, and disease in the nervous tissues produced, by morbid processes commencing in the membranes, bones and other structures by which the nerve centres or peripheral nerves are surrounded or with which they are in direct contact; by disease of the vessels with which they are supplied with blood; by alterations in the composition of the blood, the presence of poisons in the blood, the defective removal of waste products, etc.; by morbid conditions affecting the connective tissue which is so widely diffused throughout the whole nervous system; and, as I have already said, by disease or irritation of distant (peripheral) organs and parts.

Further, in some cases, but they are, comparatively speaking, rare, the lesion (morbid process) appears to commence in the nerve elements (nerve cells or nerve tubes) themselves.

The same forms of morbid process and pathological change occur in the nervous system as in the other organs and tissues of the body. The more important forms and causes of organic change are:—

(1) Malformations which interfere with the functional activity or development of the nerve centres.

(2) Inflammations and their results. Inflammation plays a very important part in the pathology of nervous diseases. In this course of lectures I shall frequently use the term sclerosis—a condition which seems in many cases to be nothing more or less than the result of a very chronic inflammatory process.

(3) Degenerative changes (fatty, pigmentary, etc.).

(4) Degenerations due to defective nerve nutrition; the conditions which are termed *secondary* degenerations are examples in point.

(5) Necrosis, destruction and death—the result of arrested or defective blood supply, hamorrhagic extravasation, intense

inflammation, severe traumatic injury, the presence of new growths, etc.

(6) New growths; they are of frequent occurrence in the brain.

Functional Derangements.—The functional or nutritive derangements and diseases of the nervous system are very common and of great importance.

As the term nutritive denotes, they are in many cases due to :—(a) diminished blood supply ; (b) a deteriorated or impure condition of the blood ; (c) the deficient removal of excretory products, the result either of venous congestion or arrested lymph flow, etc.

In some cases, these functional derangements result from peripheral irritation conveyed to the nerve centres by the afferent (sensory) nerves. Epileptiform convulsions and (reflex) paralysis may undoubtedly result from such peripheral irritation.

The nutritive and functional derangements of the brain (higher nerve centres) are of special importance. Some forms of insanity, many cases of melancholia for example, are undoubtedly due to conditions which we term functional.

That the condition of the blood exerts a most important influence upon the nutrition, development and functional activity of the nerve centres and especially the brain, is clearly shown by the extraordinary improvement in the mental condition of patients affected with sporadic cretinism and myxœdema which results from thyroid feeding. But I need hardly say that it is impossible to draw a sharp line of demarcation between functional and organic changes ; the two run insensibly one into the other ; every disturbance of function is no doubt attended with some structural change, or at all events with some change of a chemical or physical kind, which our present methods of investigation and observation are unable to reveal and detect.

Discharging and Destroying Lesions.—The functional derangements which result from nervous lesions may for the most part be divided into two great groups :—(1) In some cases, the function of the affected portion of nerve tissue is diminished or abolished ; the lesions which produce this condition are sometimes termed '*destructive*' or '*destroying*' lesions. (2) In other cases, the function of the affected part is increased or exaggerated ; the lesions which produce this form of disturbance

are sometimes termed '*irritative*' or '*discharging*' lesions. Paralysis, anæsthesia and dementia are usually the result of a destructive lesion; while convulsions, hyperæsthesia and maniacal excitement are usually due to an irritative or discharging lesion. It must, however, be remembered that in many cases the increased function which results from an irritative or discharging lesion is perverted in character. Further, it is important to note that in some cases in which the effect of the primary lesion is diminution or abolition of function, the ultimate or external result is the production of increased function. The *inhibitory* functions of the nervous system are of the greatest importance. The nervous system is, as we say, built up of a series of centres. The higher centres control the lower. Destruction of the higher (controlling) centres permits of overaction of the lower (subordinate) centres, which they (the higher centres) normally control. It is probable that in some forms of maniacal excitement the lesion is not an irritative or discharging, but a destroying, one; in other words, that the maniacal excitement is in reality due to defective or diminished action of the higher (psychical) centres, which permits of overactivity of the lower centres.

So much by way of introduction. In the lecture to-morrow we shall commence the consideration of the diseases of the spinal cord.

LECTURE II

THE DISEASES OF THE SPINAL CORD

ANATOMICAL, PHYSIOLOGICAL AND PATHOLOGICAL INTRODUCTION

THE diseases of the spinal cord are numerous; some of them are common; and since not a few of them cripple the patient or even destroy life itself, they are of great practical importance. Our knowledge of the diseases of the spinal cord is now pretty full and accurate. We know very much more about the subject than we did when I graduated five-and-twenty years ago. At that time, spinal lesions were very imperfectly differentiated; paraplegia, locomotor ataxia and spinal meningitis were almost the only affections of the spinal cord which were recognised by the general practitioner; but paraplegia is a symptom, not a disease, and in many of the cases which (on account of the muscular rigidity by which they were characterised) were at that time considered to be meningitis, we now know that the membranes of the spinal cord are not inflamed. Even in many of our large hospitals comparatively little more than this was known, although Duchenne, Lockhart Clarke, Charcot, Leyden and others, who were making a special study of the subject, had already investigated and described many of the other affections of the spinal cord with which every well-informed physician is now intimately acquainted.

The great advance in our knowledge which has taken place during the last five-and-twenty years is mainly due to an increased knowledge of the anatomy, physiology and pathology of the spinal cord; and has largely resulted from the improvements which have taken place in the methods of histological and physiological research, from the facts which have been learned by experimental investigation on living animals, and from the keenness and enthusiasm with which the clinical and

pathological features of the diseases of the nervous system have been worked out and investigated.

I cannot too strongly insist upon the fact that the key to an intelligent knowledge of the diseases of the spinal cord is an accurate knowledge of its anatomy, physiology and pathology.

It would be out of place in a course of lectures on systematic medicine to give a minute and detailed account of the anatomy, physiology and pathology of the spinal cord; I shall refer to the more important points as we go on in connection with the individual diseases. In this way, I shall be better able to indicate and emphasise the bearings of the chief anatomical, physiological and pathological facts upon the clinical phenomena. You will find that this plan is both more instructive and more interesting than that which is usually followed. But before commencing a description of the individual diseases, it will perhaps be advisable to refer to some of the fundamental and essential points. Some of them will no doubt seem to you, who are just fresh from your anatomical, physiological and pathological studies, elementary; but they are so essential for a correct understanding of the clinical side of the subject that I make no apology for bringing them before your notice. The time which I shall occupy in refreshing your memories on these fundamental points will not be misspent.

Anatomical Considerations.

Form and length.—The spinal cord is a long, comparatively thin and narrow mass of nervous tissue, roughly cylindrical in shape, which extends from the foramen magnum above to the lower border of the first lumbar vertebra below (see fig. 1).

Observe that the spinal cord does not occupy the whole length of the spinal canal. The total length of the cord is not, as a rule, more than eighteen inches. The lower part of the spinal canal is occupied by the nerve strands of the cauda equina.

Relationship to membranes, bones, etc.—The spinal cord is surrounded by the spinal membranes, bathed in the spinal fluid, held or rather swung in position by the nerve roots and ligamentum denticulatum, and protected from injury by the strong

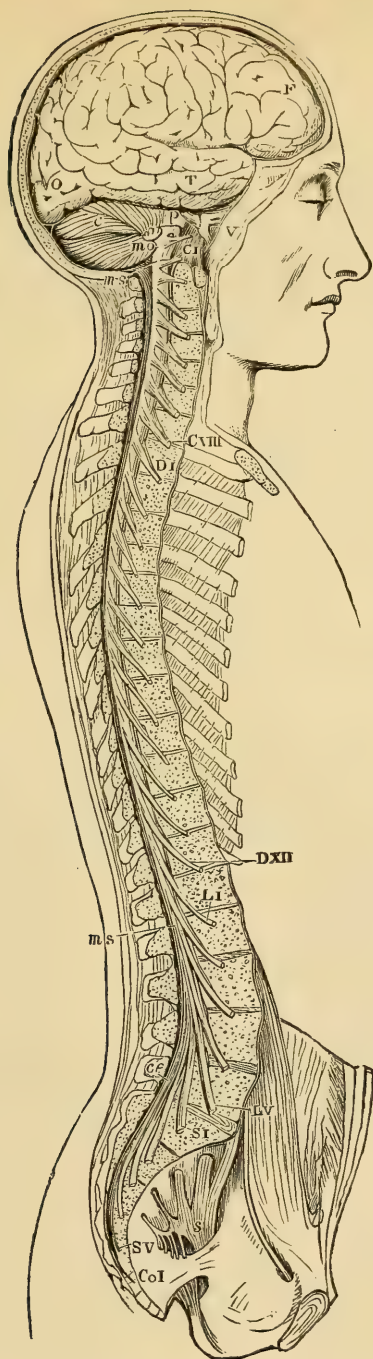


FIG. 1.—View of the cerebro-spinal axis. (After Bourguery.)

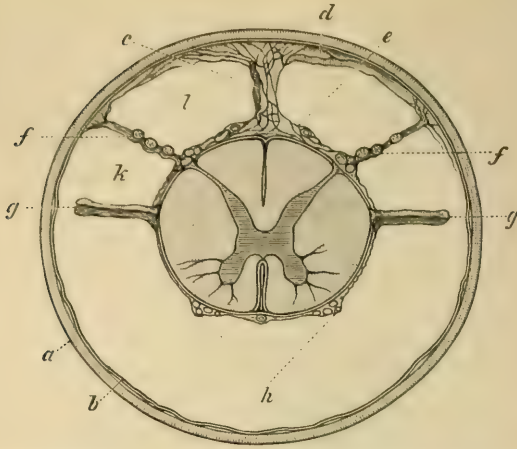


FIG. 2.—*Diagrammatic section through the Spinal Cord and its membranes in the upper dorsal region. (After Key and Retzius.) Magnified.*

a, dura mater ; *b*, arachnoid ; *c*, septum posticum ; *d*, *e*, *f*, sub-arachnoid trabeculae, those at *f*, *f*, supporting bundles of a posterior nerve-root ; *g*, ligamentum denticulatum ; *h*, sections of bundles of an anterior nerve-root ; *k*, *l*, sub-arachnoid space.



FIG. 3.—*Transverse section through the cord of a child three weeks old, with the membranes and nerve-roots in situ, showing the crossed pyramidal tracts undeveloped, i.e. faintly stained. (Stained with perosmic acid.)*

a, *a*, crossed pyramidal tracts ; *b*, divided nerve-root bundles between the pia and arachnoid ; *c*, dura mater.

bony canal in which it, and the other structures which I have just enumerated, are encased (see figs. 2 and 3).

It is important to note that the spinal cord is very securely protected from injury. Indeed, except in its upper part (where the greater flexibility of the neck renders the vertebral column more liable to dislocation, and the cord more liable to be crushed by a sudden backward twist of the neck, even when there is no fracture or dislocation), the spinal cord is one of the best and most securely protected structures in the whole body.

The spinal segment.—The spinal cord is made up of a series of segments placed one above the other; each segment comprises a portion of cord to which a pair of spinal nerves is

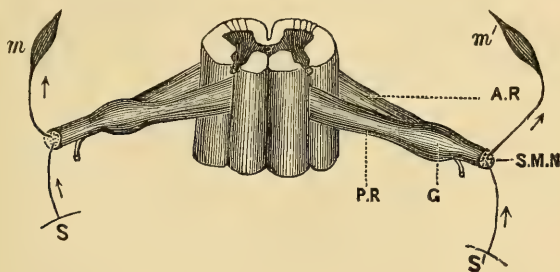


FIG. 4.—A Spinal Segment with its Muscular and Sensitive Body-Areas attached.

A.R.—Anterior root. P.R.—Posterior root. G—Ganglion on posterior root. S.M.N.—Transversely divided sensori-motor nerve trunk. *m*, *m'*—Muscular area of the segment. *S*, *S'*—Sensitive area of the segment.

attached (see fig. 4). Each segment may be viewed as a distinct spinal unit, or, to speak somewhat figuratively, as a distinct spinal cord for a definite area of the body, namely, that portion of muscle (*muscular area*) to which its anterior roots are distributed; and that portion of skin, tendon, muscle, mucous membrane, viscus, etc. (*sensitive area*) to which the fibres of its posterior roots are distributed.

The size and shape of the spinal segments vary considerably at different levels (see fig. 5). The segments composing the cervical and lumbar enlargements (those segments which are connected with the upper and lower limbs) are large, in consequence of the great size of their contained grey matter.

Speaking generally, a segment of the spinal cord may be

described as a disc of nerve tissue to which a pair of spinal nerves is attached, each nerve rising by an anterior and a posterior root (see fig. 4).

Each segment is divided into two symmetrical halves by the *anterior and posterior median fissures* (see fig. 6). The division

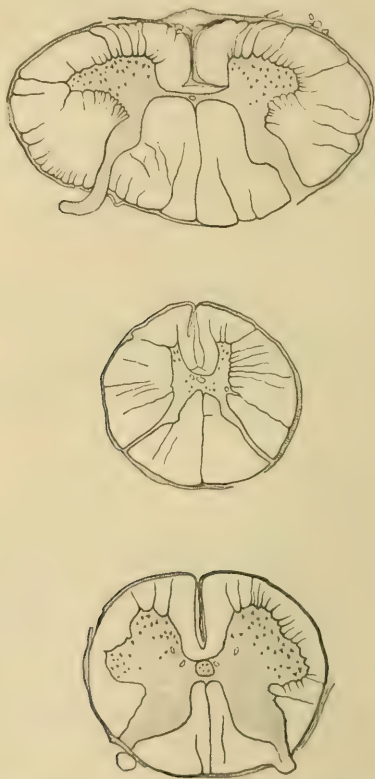


FIG. 5.—*Transverse sections through the cervical, dorsal, and lumbar regions of the Spinal Cord, showing the relative size and shape of the component parts of the transverse section.*

is not quite complete, for, towards the centre of the segment, a narrow band of nervous tissue connects the two lateral halves of which it is made up. This commissure or connective band of nervous tissue is composed partly of grey and partly of white matter. The white portion of the commissure lies in front, at the bottom of the anterior median fissure, and is termed the

anterior or white commissure. The grey portion lies behind, between the anterior white commissure and the posterior fissure or septum (for the posterior median fissure is not a fissure properly so-called, but a septum of connective tissue), and is termed the *posterior or grey commissure.* In the middle of the posterior or grey commissure, the *central canal* of the spinal cord with its beautiful lining of ciliated epithelium is placed.

Each half segment consists partly of white and partly of grey matter.

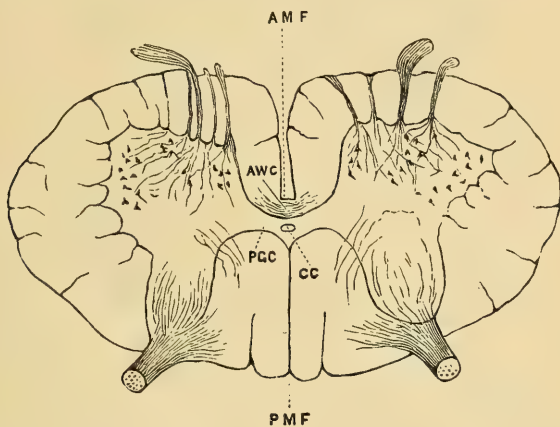


FIG. 6.—*Transverse section of a Spinal Segment in the Cervical region.*

AMF—Anterior median fissure. PMF—Posterior median fissure or septum. AWC—Anterior or white commissure. PGC—Posterior or grey commissure. CC—Central Canal.

The *grey matter* is composed of a fine nervous and connective tissue (neuroglial) matrix and a vascular network, in which the nerve cells are imbedded, and through which nerve fibres and delicate nerve fibrils run in all directions.

The grey matter (see fig. 7) consists of two extremities or horns (cornua, as they are termed) joined by an intermediate connecting portion. The *anterior horn* is much larger than the posterior, and in it are situated the large multipolar (motor) nerve cells (see fig. 8) which, as we shall presently see, are of the greatest physiological and clinical importance. The grey matter of one (say the right) half of each segment is connected with the grey matter of the other (say the left) half of that segment, by the posterior or grey commissure.

The white matter of each segment has been divided by anatomists into the following parts or columns (see fig. 9):—

1. *The posterior column*; that portion of white matter which is situated between the posterior median fissure on the one side,

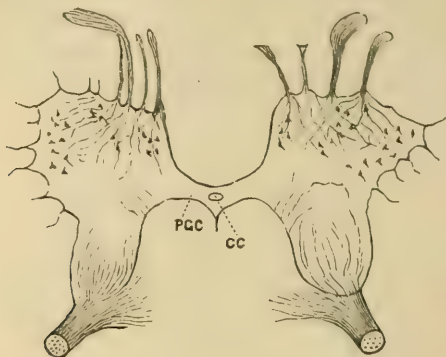


FIG. 7.—*The Grey Matter of the Spinal Segment.*
PGC—Posterior or grey commissure. CC—Central Canal.



FIG. 8.—*Nerve Cell from the Anterior Horn of the Spinal Cord.*—(After Gerlach.)
a—Axis cylinder process. b—Pigment granules in the cell.

and the posterior horn of grey matter and the posterior nerve root, on the other.

2. *The lateral column*; that portion of white matter which is situated between the posterior and the anterior nerve roots.

3. *The anterior column*; that portion of white matter which is situated between the anterior median fissure and the lateral column, i.e. the innermost (most laterally placed) of the anterior root bundles.

In each of these columns of white matter there are situated certain tracts or areas of nerve fibres, each of which has a definite and distinct physiological function. The exact position and function of the more important of these physiological areas or

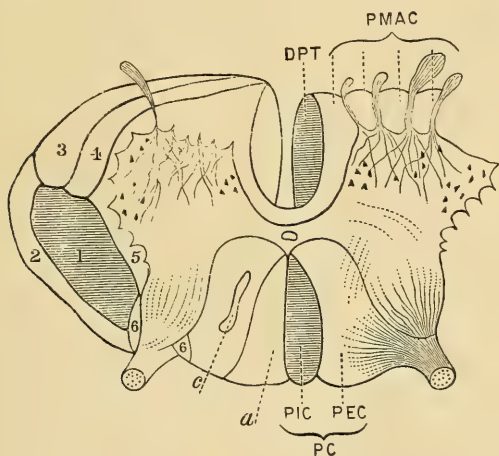


FIG. 9.—*Diagrammatic Section of a Spinal Segment showing the Physiological division of the White Matter.*

The lateral column of the right half of the segment has been cut away. PIC—Postero-internal column. PEC—Postero-external column. DPT—Direct pyramidal tract. PMAC—Principal mass of the anterior column. 1. Crossed pyramidal tract. 2. Direct cerebellar tract. 3. Antero-lateral ascending tract (Gowers' tract). 4. Antero-lateral descending tract. 5. Internal boundary layer of the grey substance. 6, 6—Lissauer's tract. *a*—Postero-internal column. *c*—Comma-shaped tract.

tracts (those which, in the present state of our knowledge, are of chief practical and clinical importance) will be particularly described when I come to speak of the individual diseases of the cord in detail.

You will observe that, in the spinal cord, the grey matter is placed in the centre and completely surrounded by white matter, the very reverse of the arrangement which obtains in the brain; for in the cerebrum (and cerebellum) the grey matter is for the most part spread, in the form of a comparatively thin layer, over

the surface of the great masses of white matter of which the cerebral hemispheres are largely composed. This difference in the relative position of the grey and white matter, in the brain and spinal cord respectively, is of practical (clinical) importance, as we shall see when I come to describe some of the individual diseases, more especially the inflammations of the membranes and the other surface lesions.

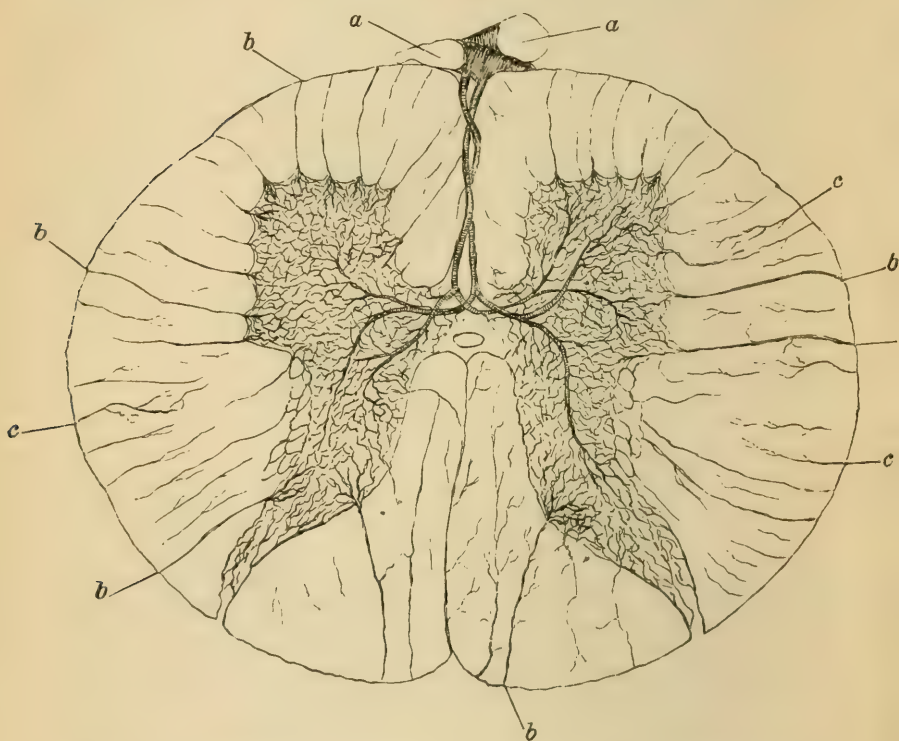


FIG. 10.—*Distribution of the blood-vessels in the Spinal Cord.*—(After Adamkiewics.)

Blood supply.—The manner in which the spinal cord is supplied with blood has also a very important practical bearing. The main arterial supply is derived from the anterior and posterior spinal arteries (branches of the vertebrals), which, in comparison to their great length, are very slender vessels (see fig. 10). In addition, a minute arterial twig passes to the cord along

each nerve root; these arteries are branches of the intercostal, lumbar, ileo-lumbar and sacral arteries.

The arteries which supply the lower end of the cord, both those which reach it from above (the anterior and posterior spinal arteries) and those which reach it from below (the branches which pass to it along the strands of the cauda equina) are very long and slender vessels; and, since the blood flow meets with considerable mechanical obstruction, in passing through such long and slender elastic vessels, the arterial blood supply to the lower end of the cord is, comparatively speaking, with difficulty maintained. This is perhaps (as the late Dr. Moxon was the first to suggest) the reason why softenings and some other lesions are, comparatively speaking, more common in the lumbar and lower dorsal than in the cervical and upper dorsal regions of the cord.

Physiological Considerations.

The physiology of the spinal cord, and its functional relations to the brain above, to the periphery below, and of its own individual parts to one another, is much more easily understood if we get into the habit of regarding each segment as:—

1. *A conducting medium*, so to speak (see fig. 11), *through which* the great nerve tracts (motor, sensory, controlling, etc.) pass, which place the brain in connection with all the other segments situated below, or, as comparative anatomists say, posterior to, it (i.e. the segment we are dealing with).

2. *A centre* (see fig. 12), (a) *from* which efferent motor, reflex, vaso-motor, and trophic influences are distributed *to*; and (b) *to* which afferent impressions (concerned in the production of sensations, reflex movements, and the co-ordination of movement) proceed *from* a limited area of the body, i.e. that portion of the body with which the pair of spinal nerves belonging to the segment is connected.

In fig. 13, the spinal segment is represented in both of these capacities, i.e. as a conducting medium and a spinal centre.

The special functions of the different parts of the spinal segment will be considered in detail in connection with the individual diseases and lesions.

Relationship of different segments to one another.—Though each half-segment is to a large extent functionally independent

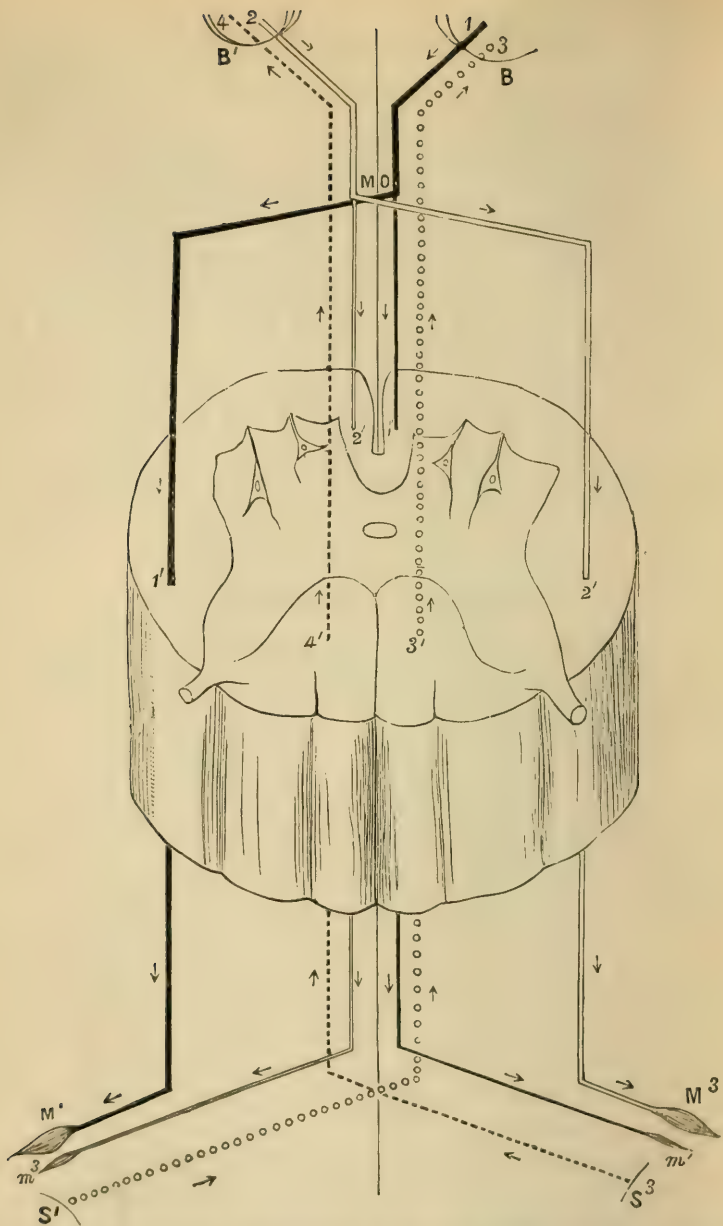


FIG. 11.—Diagrammatic representation of the Spinal Segment as a conducting medium, showing the passage of the main motor and sensory tracts through it. The muscular and sensory areas of the segment have been cut away.

B, Right, and B', Left hemispheres of the brain. MO, Lower end of medulla oblongata. 1, 1', Pyramidal (motor) tract from the right hemisphere passing through the segment in the lateral column of the opposite, and in the anterior column of the same side. 2, 2', Pyramidal tract from the left hemisphere. 3', 3 and 4', 4, Main sensory tracts passing up to the brain in the postero-internal columns.

The arrows indicate the direction of the nerve impulses.

of its fellow on the opposite side—an independence which is more marked in the segments composing the cervical enlargement, in order to provide for the highly specialised movements of the upper limbs—each half-segment is in intimate physiological relationship with its fellow on the opposite side. Further, adjacent segments are not only structurally, but functionally,

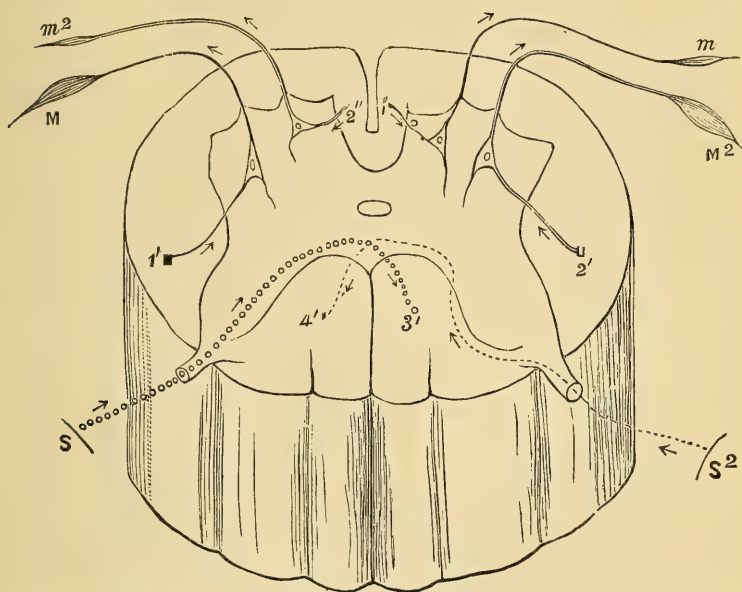


FIG. 12.—*Diagrammatic representation of the Spinal Segment as a Spinal Centre.*

1' 2' Motor fibres proceeding from the left crossed and direct pyramidal tracts, to supply the muscular area (M, m^2) of the left lateral half of the segment. 2' 1' Motor fibres proceeding from the right crossed and direct pyramidal tracts to supply the muscular area (M^2 , m) of the right lateral half of the segment. S, Sensory fibre proceeding from the left sensitive area of the segment to join the main sensory tract (3') in the right postero-internal column. S², Sensory fibre proceeding from the right sensitive area of the segment to join the main sensory tract (4') in the left postero-internal column.

The arrows indicate the direction of the nerve 'impulses.'

connected and related to one another. Such a combination is required (to give one example only) for the conduction and grouping of motor impulses. But in addition to these, which we may term the more highly specialised commissural connections, every segment is connected with every other segment by means of the vast nerve network of delicate nerve fibrils which constitute such

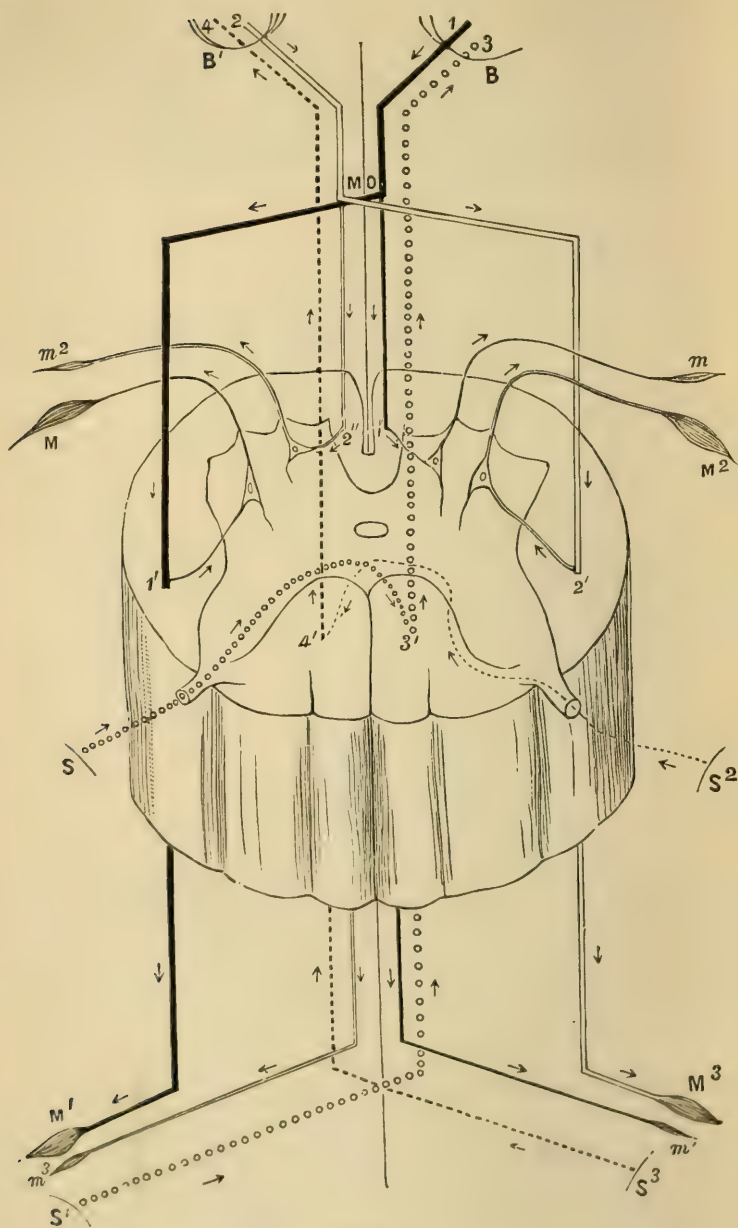


FIG. 13.

a large and important part of the grey matter. This connection is of great practical importance, for, as we shall afterwards see, when a nerve current (to speak in popular language) is 'blocked' by a lesion and prevented from flowing in the direction in which it is normally in the habit of flowing, it tends to make its way in some other direction, in other words, to evade the block by passing through some side or collateral channel. In some cases, the position of the lesion (the block) prevents the possibility of such a bye-passage. Section of a peripheral nerve or of its posterior root will, of course, absolutely arrest ('block') all afferent (incoming) impressions and will absolutely prevent them reaching the spinal cord at all. But if the lesion is situated in the grey matter, the nerve current, or nerve vibration to speak more accurately, although unable to make its way along the normal and accustomed channel, may pass through adjacent portions of the grey matter which remain open (are undestroyed), in an unaccustomed (abnormal) direction. These bye-paths are of

DESCRIPTION OF FIG. 13.

Diagrammatic representation of the Spinal Segment as a Spinal Centre and Conducting Medium.

B, Right, and B', Left, hemispheres of the brain. MO—Lower end of medulla oblongata.

1, Motor tract from the right hemisphere. At MO it divides. The larger subdivision decussates, passes down the lateral column of the opposite side of the cord, and supplies the muscular fibres M and M' on the left side of the body. At 1' the supply to M. is given off. The smaller subdivision does not decussate, but passes down the anterior column, and supplies the muscles *m* and *m'* on the same (right) side of the body.

2, The motor tract from the left hemisphere. It supplies the muscles M² and M³ on the right side of the body, and the muscles m² and m³ on the left side of the body.

S, S', Sensitive areas on the left side of the body. 3', 3 the main sensory tract from the left side of the body; it passes up the right (opposite) side of the cord in the (?) postero-internal column, and proceeds to the right hemisphere of the brain.

S², S³, Sensitive areas of the right side of the body. 4', 4, The main sensory tract from the right side of the body, proceeding up the left side of the cord to the left hemisphere of the brain.

Note to Figs. 11, 12, and 13.

Recent researches have shown that some of the fibres of the pyramidal tract pass down (undecussated) in the lateral column (crossed pyramidal tract) on the same side of the cord.

It must be remembered that the direct pyramidal tract terminates about the middle of the dorsal region, and is not therefore seen in the lower dorsal and lumbar segments.

The exact course of the sensory tracts in the cord is yet undetermined. Some authorities think that the decussation is total, and that all the sensory fibres pass up the posterior columns; others, that the sensory decussation is partial, some fibres passing up the posterior, others up the lateral column.

great importance for the purposes of compensation. Further, they enable us to understand the manner in which certain diseased phenomena (certain symptoms which are met with in cases of disease) are produced—phenomena which it would otherwise be difficult or impossible to explain.

Pathological Considerations.

In some of the cases in which the functions of the spinal cord are deranged, the lesion begins in the tissues of the cord; in others, in the structures which surround it (the membranes, bones, etc.). It is convenient to separate these two groups, and to term them *intra-* and *extra-*medullary lesions respectively.

Intra-medullary lesions.—In some of the diseases of the spinal cord in which the lesion commences in the cord (is intra-medullary), the lesion is more or less sharply defined and restricted to a definite physiological area or tract (to definite systems of fibres or areas of grey matter). These we term '*system*' lesions, or '*system*' diseases. In others, the morbid process or lesion is not so defined or restricted; it involves at hap-hazard, as it were, a greater or smaller area of the transverse section of the cord, irrespective of the physiological areas and tracts to which I have just referred. To these, I have applied the term '*indiscriminate*' lesions.

It not unfrequently happens that these two forms of lesion are combined. Thus, a morbid process, which is at first restricted to a definite physiological area or tract, which, in other words, is a system lesion, may extend to adjacent areas or parts and become to some extent indiscriminate. The reverse condition, in which an indiscriminate lesion is followed by a system lesion, is, however, far more common; a transverse myelitis, for example, produces secondary descending and ascending degenerations.

The System Diseases of the Spinal Cord.—The great characteristic of a system disease of the spinal cord is, as I have previously stated, the sharp limitation of the lesion to a definite physiological area or tract. A system lesion usually extends through several segments, sometimes indeed through the whole length of the cord, and is generally bilateral and symmetrical.

The system lesions of the spinal cord are either *primary*, i.e. arising independently of any previous lesion; or *secondary*, i.e. resulting from some previous morbid condition in the brain, spinal cord itself, or the posterior nerve roots.

The secondary system lesions are usually termed *secondary degenerations*, descending or ascending as the case may be. In the case of these secondary degenerations, the lesion is a nutritive change due to impairment or arrest of the nutritive force, which the trophic centre exercises on the nerve fibres of the tract which is affected; hence the term *nutritive* or *trophic* degenerations, which I am in the habit of using.

The chief physiological areas or tracts in the transverse section of the cord which are liable to be affected by primary system lesions are:—1. The anterior cornu; 2. The crossed pyramidal tract; 3. The postero-external column. (See fig. 14.)

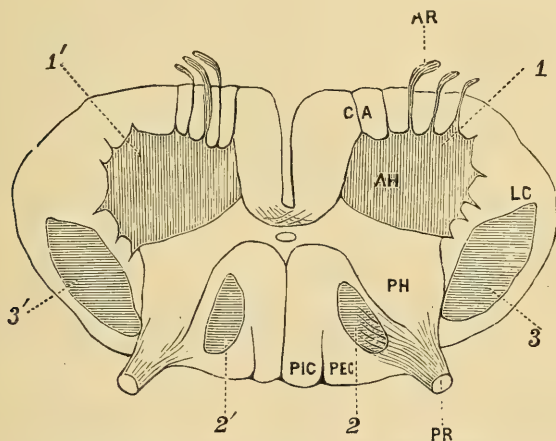


FIG. 14.—Diagram showing the primary system lesions of the Spinal Cord.

1, 1', Region of the anterior horn. 2, 2', Region of the postero-external column. 3, 3', Region of the crossed pyramidal tract.

The areas of the transverse section which are chiefly affected by secondary system lesions (secondary nutritive or trophic degenerations) are:—1. The crossed pyramidal tract; 2. The direct pyramidal tract; 3. The postero-internal column; and 4. The direct cerebellar tract. (See fig. 15.)

In the majority of cases, one system only is involved; but, in some instances, two physiological areas of tracts in the same

half-segment are affected. The condition termed postero-lateral sclerosis is an example in point.

The Indiscriminate Lesions.—The great characteristic of an indiscriminate lesion is, as I have already stated, the fact that it is not of necessity limited to any particular physiological area or tract, but that it may affect at hap-hazard, as it were, any part of the transverse section of the cord.

It may, of course, happen that an indiscriminate lesion is accidentally, so to speak, limited to a given physiological area or

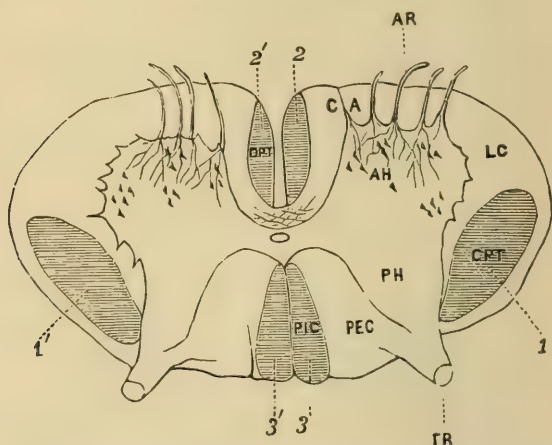


FIG. 15. --Diagram showing the secondary system lesions of the Spinal Cord.

1, 1', Region of the crossed pyramidal tract. 2, 2', Region of the direct pyramidal tract. 3, 3', Region of the postero-internal column. 4, The system lesions of the direct cerebellar tract and of Gowers' tract are not represented in the figure.

tract. In such cases, the symptoms (the results of the lesion) may exactly correspond to those which are caused by a system lesion of the same area or tract; the two conditions in fact may be indistinguishable. It is probable that in some cases of poliomyelitis anterior acuta in the adult, the lesion is of this nature,—an indiscriminate inflammation which is accidentally limited to the anterior horn of grey matter, and which, in respect to its causation (etiology), is distinct from the special form of inflammation, which is the pathological basis of the system disease poliomyelitis anterior acuta in the child.

The vertical extent of indiscriminate lesions is, as a rule, small; indeed, the lesion may be limited (in its vertical extent,

i.e. from above downwards) to a single segment. But, in many cases in which the lesion is indiscriminate, many separate foci of disease (many separate lesions) are scattered throughout the cord. Such a condition does not constitute an exception to the general rule, for the vertical extent of each individual patch of lesion is, as a rule, small; in many cases of this kind, the individual patches are separated from one another by healthy portions of nerve tissue, they are not anatomically and structurally continuous. It must of course be remembered, that secondary degenerations (ascending and descending) often result from indiscriminate lesions; this degenerative process (this secondary degeneration), which may extend through long areas of the cord, must not be confounded with the primary (indiscriminate) lesion.

The more important indiscriminate lesions are:—myelitis, simple non-inflammatory softening, cerebro-spinal sclerosis, intra-medullary hæmorrhage, and intra-medullary tumours.

I may take this opportunity of saying that hæmorrhagic extravasation and embolic plugging of the arteries—lesions which are so common and so important in the brain—are extremely rare in the spinal cord. This difference is due to the facts, that the blood pressure in the arteries of the spinal cord is, compared to that of the brain, very low; and that the arteries of the cord arise at an angle from the vertebrae and are, in comparison with those of the brain, very small. Further, I may say that tumours both in the substance and on the surface of the spinal cord—intra- and extra-medullary tumours respectively—are also very rare in comparison with tumours (intra- and extra-medullary) of the brain.

The Extra-medullary Lesions.—The chief affections comprised in this group are:—Pott's disease of the vertebræ, traumatic injuries such as fractures or dislocations of the vertebræ, wounds and contusions of the cord, spinal meningitis, tumours in the spinal canal springing from the bones, membranes or nerve roots, and hæmorrhagic extravasations into the spinal canal. Many of these lesions, such, for example, as extra-medullary tumours and extra-medullary hæmorrhage (if we except the hæmorrhages which are associated with, and which result from, injuries—fractures and dislocations of the vertebræ), are very rare. Further inflammation of the spinal membranes is, in comparison with

inflammation of the cerebral membranes, a rare condition; and when it does occur it is relatively unimportant; for, in many of the cases in which the spinal membranes are inflamed, the cerebral membranes are also affected, and in such cases the inflammation of the spinal membranes is usually, both pathologically and clinically, subordinate and secondary to the inflammation of the cerebral membranes, and, comparatively speaking, slight. Again, it must be remembered that the spinal cord is far better protected than the brain from external injury; and that fractures and dislocations of the spine (though these lesions are common enough) are with difficulty produced; while fractures of the skull are, comparatively speaking, readily produced, and are relatively much more frequent.

Lastly, I should state that a true *concussion of the spinal cord* (concussion irrespective of a definite lesion such as a fracture or dislocation of the vertebral column), is, except perhaps in pitmen, a rare condition; while concussion of the brain (without fracture of the cranial bones) is very readily produced and of every-day occurrence.

Malformations of the spinal cord are very uncommon and unimportant, though the condition which is termed syringomyelia, which consists in the presence of cavities in the spinal cord, and which in many cases seems to be the result of congenital malformation or defective development, is of great interest and of considerable practical importance.

We are now in a position to consider the individual diseases of the spinal cord in detail. I shall commence with the intra-medullary lesions. We will first consider the primary system lesions, for in trying to master any difficult and complicated subject it is always advisable to begin with the simple and proceed to the complex. If you succeed in getting a thorough grasp of the symptoms which result from the primary system lesions—the diseases which result from lesions of localised areas and definite physiological tracts—you will find it comparatively speaking an easy matter to understand the symptoms which characterise the more complex indiscriminate and extra-medullary lesions.

LECTURE III

POLIOMYELITIS ANTERIOR ACUTA

Synonyms.—Various synonyms have been given to this disease; the more important are:—‘*Acute Atrophic Spinal Paralysis*,’ ‘*Infantile Paralysis*,’ ‘*The Essential Paralysis of Children*,’ and ‘*Regressive Paralysis*.’

As the term poliomyelitis anterior acuta denotes, the disease is due to an acute inflammation of the anterior horn of grey matter. It is very much more common in children than in adults, hence the synonyms ‘infantile paralysis’ and ‘the essential paralysis of children.’ But neither of these are good terms, and that for two reasons; in the first place, this is by no means the only disease which produces paralysis in children—the term ‘infantile paralysis’ should include every form of paralysis which occurs in the child; and, in the second place, poliomyelitis anterior acuta is not confined to childhood—it is sometimes, though comparatively speaking rarely, met with in the adult.

Morbid Anatomy.—The opportunity of observing the condition of the cord in an early stage rarely occurs, for the disease is very seldom fatal. A few cases have, however, been examined; the earliest with which I am acquainted is reported by my friend Dr. David Drummond of Newcastle: in that case, the child died a few hours after the commencement of the attack, and the changes in the cord were undoubtedly inflammatory in character. From this and other cases it would appear, that the morbid changes in this disease are very similar to those which are met with in the ordinary form of myelitis—a common condition which is often fatal.

Now, in the common form of inflammation of the spinal cord (ordinary transverse myelitis) the same changes occur which are characteristic of inflammation in every vascular organ, viz., congestion and stasis, inflammatory exudation and extravasation,

proliferation of the tissue elements, and, in cases in which the inflammation is acute and very severe and the inflamed tissues soft and delicate—and the grey matter of the spinal cord is very delicate and easily destroyed—it may be extensive destruction of the inflamed part.

The grey matter of a cord which is acutely inflamed usually looks redder than normal; but this is not always the case; even after transverse section, the cord may appear normal to the naked eye. Microscopic examination shows that the vessels are engorged and that the inflamed tissues are infiltrated with leucocytes and exudation products. Small punctiform hæmorrhages are often, and larger hæmorrhages are occasionally, present.

In the slighter cases, the nerve elements are not extensively destroyed; but in cases of average severity there is usually some destruction of tissue. So far as I know, swelling and enlargement of the nerve cells and axis-cylinders, which are such conspicuous and characteristic features of the ordinary form of inflammation (myelitis), have rarely if ever been observed in poliomyelitis anterior acuta; but this is perhaps due to the fact that very few cases have been examined during the acute stage of the disease.

In more severe cases, the destruction of tissue is much greater. The anterior horn may be ploughed up, as it were, by the extravasation and exudation.

In some cases, the onset of the paralysis is so sudden as to suggest a primary hæmorrhage rather than an inflammation. But primary hæmorrhage in the spinal cord is exceedingly rare; and in those cases of poliomyelitis anterior acuta in which the onset is so sudden as to suggest a hæmorrhage, it is probable that the extravasation of blood is the result and not the cause of the inflammatory changes, which, up to the onset of the hæmorrhage, were so slight that they had not produced any marked symptoms.

Some authorities have supposed that the morbid change commences in, or is entirely confined to, the multipolar nerve cells of the anterior horn. In fact, an acute atrophy or parenchymatous inflammation of the ganglionic cells, characterised by granular pigmentation followed by disappearance of the cell processes and ultimately resulting in complete atrophy of the cells themselves, has been described. But the occurrence of such a lesion is very doubtful. There seems strong reason to suppose

that the inflammatory condition starts around the vessels, and that it is probably due to the presence of some toxic irritant in the blood. In quite recent cases, the blood-vessels proceeding to the inflamed area are sometimes plugged or thrombosed.



FIG. 16.—*Transverse section of the Spinal Cord in the Lumbar region, from a case of Infantile Paralysis (carmine and dammar), magnified about 10 diameters.*

The membranes are still *in situ*. Numerous transversely divided nerve roots (b) are seen between the dura and the pia mater. The process of membranes, which passes into the anterior median fissure, is slightly displaced; a few of the transversely divided nerve roots (c) have escaped outside the dura in the process of mounting.

Nearly all the multipolar nerve cells of the left anterior cornu have disappeared. The inner group still remains. Compare with fig. 17. Some of the anterior root bundles (a) can be seen to be degenerated even with this low power. The central canal is of large size; under a high power it is seen to be beautifully healthy.

Remember, then, that in those cases in which the inflammation is slight, there may be merely inflammatory exudation and œdema, but little destruction of tissue; while in severe

cases, the grey matter of the anterior horn may be extensively destroyed.

After the inflammation has continued for some little time, the acute changes subside and the inflammatory products are absorbed. The ultimate result is, in the slighter cases, destruction of some of the nerve cells and nerve fibres and the production of some cicatricial thickening, sclerosis, as we term it, of the affected part of the anterior horn of grey matter; and in



FIG. 17.—*Transverse section through the Lumbar region of the Spinal Cord of a Child, showing the normal appearance of the Anterior Cornua (carmine and dammar) magnified about 10 diameters.*

Numerous multipolar nerve cells are seen in the anterior cornua. The central canal is double. This condition is a rare anatomical variation of no practical importance.

the more severe cases, the destruction of a large number, in fact it may be of all, of the nerve cells in the inflamed area, extensive sclerosis, cicatricial shrinking of the anterior horn and of the adjacent white columns; and, in some cases, though this is rare, the formation of a cavity surrounded by cicatricial fibrous tissue in the anterior horn of grey matter.

This drawing (see fig. 16), which represents a transverse section of the lumbar region of the cord, shows the appearances seven weeks after the onset of the inflammation. The patient was attacked with poliomyelitis anterior acuta on the 21st March,

and died from diphtheria on the 9th May, 1879. The nerve cells in the left anterior horn have almost entirely disappeared.



FIG. 18.—*Transverse section through the Anterior Horn of grey matter in a case of Infantile Paralysis (osmic acid and farrant, low power).*

Nearly all the nerve cells have disappeared. Their place is taken by fatty particles which can be easily seen even with this low power.



FIG. 19.—*Portion of the Anterior Horn of grey matter represented in fig. 18, showing the minute structure of the lesion (osmic acid and farrant).*

The dark particles *a* seen in fig. 18 are fatty globules stained with osmic acid. They are situated in spaces which have formerly contained nerve cells. A small vessel *b* is seen at the upper part of the section—some fatty globules adhere to its outer coat.

Under a higher power (see figs. 18 and 19, which represent a portion of the same section more highly magnified), fatty granules and globules of oil may, with suitable staining (osmic acid), be seen in the position of some of the nerve cells which have been destroyed.

In old-standing cases, in which the attack was a severe one, the affected anterior horn may be much shrunken (see fig. 20). It occasionally, though rarely, happens that in cases of this kind

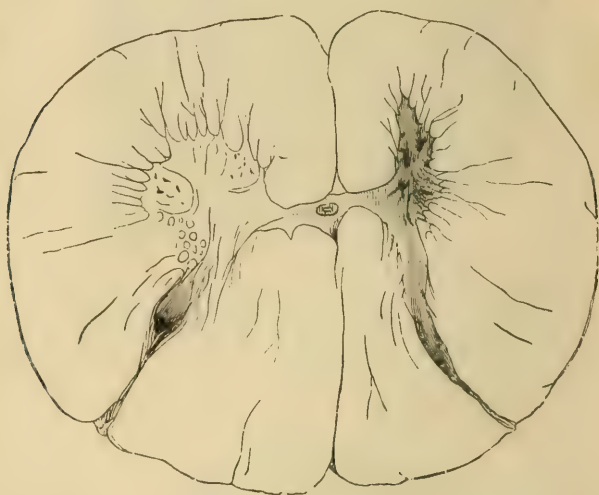


FIG. 20.—*Transverse section of the Cervical region of the Spinal Cord in a case of old poliomyelitis anterior acuta.*—(After Charcot.)

The patient, a woman *æt.* 50, died in the Salpêtrière from *General Paralysis of the Insane*. The right upper extremity had been affected in childhood with poliomyelitis anterior acuta. The right anterior cornu was cirrlosed, and the white columns of the corresponding side atrophied.

an actual cavity or cyst surrounded by a ring of sclerosed and cicatricial tissue is found in the anterior horn at the seat of maximum intensity of the lesion. (See fig. 21.)

The lesion, then, which is the pathological substratum of poliomyelitis anterior acuta is an acute inflammation of the anterior horn of grey matter. The intensity of the inflammation varies very greatly in different cases; but in almost every case, however slight the inflammation may happen to be, there is some permanent damage, that is to say, some destruction of the

nerve cells and fine motor fibres which are the active constituents of the anterior horn of grey matter.

In the great majority of cases, the inflammation is, practically speaking, limited to the anterior horn of grey matter; it is for this reason that we place poliomyelitis anterior acuta under the system diseases. The limitation of the lesion to the region of the anterior horn has, as we shall presently see, a very important bearing on the symptomatology of the disease. But although



FIG. 21.—*Transverse section through the Cervical region of the Spinal Cord in an old-standing case of poliomyelitis anterior acuta, showing a cavity (A) in the right anterior horn of grey matter. The adjacent part of the lateral column (B) is sclerosed; and the anterior and lateral columns are markedly wasted.*

the lesion is practically limited to the region of the anterior horn, it is certain that when the inflammation is severe, the adjacent parts of the cord are not unfrequently affected in some degree. This is, of course, only what one would expect. In some cases, leucocytes are scattered throughout the grey matter. In some cases, the inflammation has evidently extended to the posterior horn; in others, the adjacent parts of the antero-lateral columns are involved, but usually only in a slight degree. In old-standing cases, it is not at all uncommon to find the connective tissue trabeculae in those parts of the anterior and lateral columns which are adjacent to the anterior horn, thickened and sclerosed.

Further, it must be remembered that in some cases microscopic examination shows that the lesion is more extensively

diffused through the grey matter of the anterior horns than the clinical symptoms during life seem to suggest. In some cases, for example, in which the lesion is severe in the lumbar enlargement, evidences of slight inflammation may be detected on microscopical examination after death in the cervical and dorsal regions. Again, the clinical course of some cases seems to show that the peripheral nerves are occasionally inflamed; and a celebrated German observer, Leyden, has advanced the view that the cord changes are in some cases secondary to a neuritis of the peripheral nerves. I am not prepared to deny the possi-

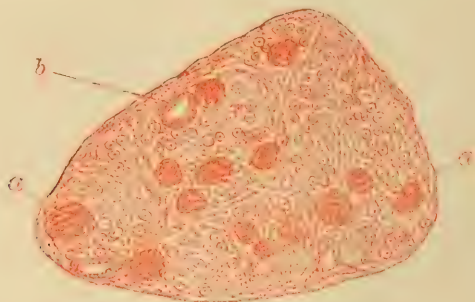


FIG. 22.—*Transverse section of an Anterior Nerve Root from a case of Infantile Paralysis, showing marked degeneration. The preparation is one of the anterior nerve root, c, seen in Fig. 16, more highly magnified. $\times 200$ diameters.*

Many of the nerve tubes have disappeared, and are replaced by masses of connective tissue, *a, a*, which stain deeply with carmine; *b*, transversely divided blood vessel.

bility of this, but it seems to me a most unlikely and improbable explanation in the great majority of cases.

Remember, then, that in most cases the inflammation is practically (for clinical purposes) limited to the anterior horn of grey matter, though exceptions to this general rule occasionally occur.

One point in the morbid anatomy of the disease still remains to be mentioned. The anterior nerve rootlets, which pass out of the cord through the anterior column, and the anterior nerve roots are always affected in some degree. In severe cases, they are extensively degenerated and atrophied (see fig. 22). At first sight, this may appear to be an exception to the statement on which I have laid so much stress, that the inflammation is confined to the grey matter of the anterior horn. But it is not

so; for the changes in the anterior nerve rootlets and in the anterior roots are not the *direct* result of the acute inflammatory process; they are due to a process of secondary or nutritive (trophic) degeneration, as I prefer to term it; and this leads me to speak of the pathological physiology of the disease.

Pathological Physiology.—What effects would you expect

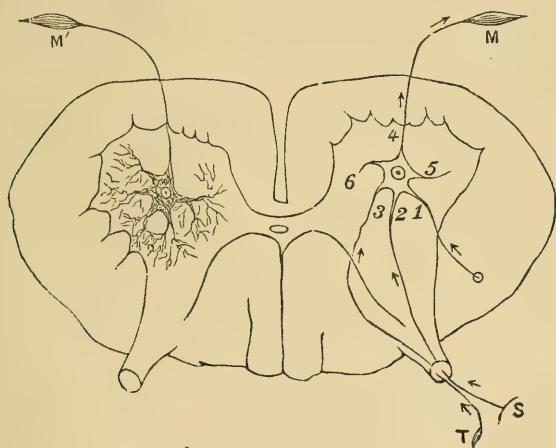


FIG. 23.—*Diagrammatic representation of the connections of the motor nerve cells of the anterior cornu.*

1. Branch which conveys to the cell, voluntary-motor and controlling-reflex impulses. 2. Branch which conducts to the cell the superficial reflexes. 3. Branch which conducts to the cell the deep reflexes. 4. Axis-cylinder process, which transmits voluntary-motor, reflex-motor, and trophic impulses, from the cell to the muscle M. 5. Branch which places the cell in communication with other cells in other segments. 6. Branch which places the cell in connection with other cells in the opposite anterior horn of the same segment. On the left side the division of the cell processes, which terminate in Gerlach's nerve network, is shown.

this lesion (this acute inflammation of the anterior horn of grey matter) to produce? What disturbances of function will it be likely to cause?

In order to answer these questions, it is essential to remember that the anterior horn of grey matter is a strictly motor area. It consists of a neuroglial basis and vascular network, in the meshes of which the large multipolar (motor) nerve cells (see figs. 23 and 24) are situated, in which motor nerve fibres and delicate nerve fibrils ramify in all directions, and through which

the axis-cylinder processes pass outwards to reach the surface of the cord and the anterior nerve roots, with the fibres of which they are directly continuous.

The multipolar nerve cells are the active and most important constituents of the anterior horn of grey matter. *To* these motor nerve cells, voluntary-motor impulses proceed from the brain above, through the fibres of the crossed pyramidal tracts. *To* these multipolar nerve cells reflex impulses also pass from the periphery, *per* the fibres of the posterior nerve roots (see fig. 23). *From* these multipolar nerve cells, voluntary-motor and reflex-motor impulses pass out to the muscles, through the axis-cylinder processes, anterior roots and motor fibres of the peripheral spinal nerves, which are direct continuations of the axis-cylinder processes.

Recent researches seem to show that the fibres of the crossed pyramidal tracts are not *directly* connected, as was at one time supposed, with the multipolar nerve cells of the anterior horn. It is now believed that the connection is not a continuous one. The fibres of the crossed pyramidal tract, after entering the grey matter of the anterior horn, divide and subdivide into fine branches (fibrils), which, it is now believed, terminate in rounded or bulbed ends. The fibres of the multipolar nerve cells (with the important exception of the axis-cylinder process which does not divide) branch and divide in the grey matter into a number of fine fibres and fibrils which, like the fine fibrils of the crossed pyramidal tract, terminate in rounded or bulbed ends. Recent observations seem to show that the two sets of fine fibres, with their terminal rounded or bulbed ends, come into the closest juxtaposition, but are not directly continuous one with another (see fig. 24).

The *first* effect, then, of an inflammation which is limited to the anterior horn of grey matter will necessarily be to interrupt the function of the motor nerve cells and the motor fibres and fibrils which are its active constituents; in other words, to produce paralysis in the muscular fibres with which the nerve cells and nerve fibres (the function of which is interrupted) are connected.

The inflammation varies, as we have seen, in intensity; and the paralysis may be due to different causes. In some cases, it is merely the result of inhibited or arrested function, due to the

'shock' which attends the onset of a severe inflammation or to the pressure which the inflammatory products exercise on the nerve elements. In others, it is due to destruction of the nerve elements. In the former case, the paralysis is merely temporary; in the latter it may be permanent. Complete destruction of a large number of nerve cells will produce permanent paralysis; for, so far as we know, nerve cells which are once completely destroyed are never restored. Destruction of axis-cylinders and fine motor nerve fibrils will not produce permanent paralysis, for, provided that the nerve cells connected with the destroyed fibres and fibrils remain healthy, the affected (destroyed) fibres and fibrils are without doubt in many cases restored.

The *first* effect, then, of an inflammation limited to the anterior horn of grey matter is to produce paralysis of the muscular fibres with which the nerve cells and nerve fibres of the affected area of grey matter are directly connected.

The *second* effect is to interrupt or permanently abolish those reflex impulses which pass through the affected area of grey matter.

The multipolar nerve cells are the reflex centres for the muscle areas (portions of muscular tissue) with which their axis-cylinder processes are directly connected (see fig. 23). Like the paralysis, the abolition of the reflex movements is in some (the slight) cases temporary, and in others (the severe cases) permanent.

The *third* effect is to produce certain trophic alterations (degeneration and atrophy) in the motor nerves and muscular fibres with which the affected parts of the anterior horns are directly connected.

The multipolar nerve cells of the anterior horn of grey matter are the trophic centres for:—(a) the axis-cylinder processes which pass out from them and for the fibres of the anterior (motor) nerve roots; (b) the motor fibres of the peripheral nerves; (c) the motor nerve-endings in the muscles (all of which are direct continuations of the axis-cylinder processes); and (d) the muscular fibres in which those motor nerve-endings terminate. The nutrition of the long but extremely delicate neuro-motor nerve tract, which extends from the multipolar nerve cell to (and including) the muscular fibre which that nerve cell innervates (see fig. 24), depends upon the healthy functional activity of this multipolar nerve cell. When the multipolar nerve cell is destroyed or its

trophic function seriously disturbed, degenerative changes immediately begin to take place in the long neuro-motor tract (axis-cylinder process, anterior root-fibres, etc.) which passes off from it. This process of degeneration is attended with definite histological changes to which I shall presently refer in more de-

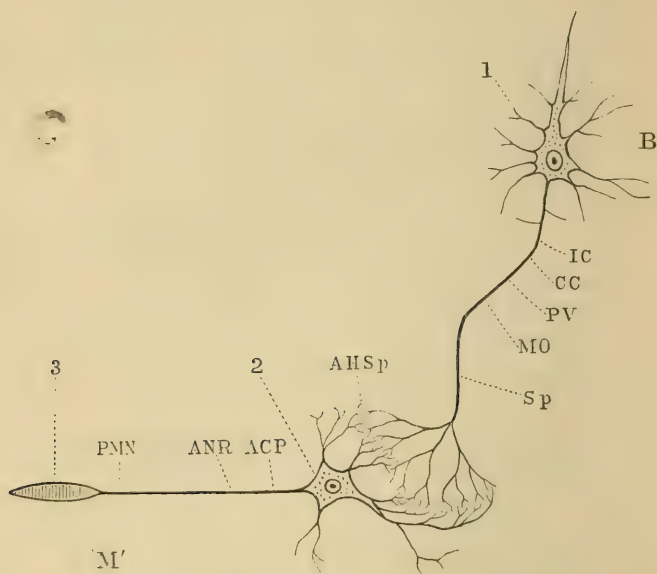


FIG. 24.—*Plan of the motor nerve apparatus.*

Figure 1 points to a large pyramidal nerve cell, in the motor area of the right hemisphere of the brain (B), from which a basal axis-cylinder process passes, as a fibre of the main pyramidal tract, down to the spinal cord, through the internal capsule (IC), crus cerebri (cc), pons Varolii (pV), medulla oblongata (Mo), and spinal cord (sp). In the grey matter of the spinal cord, this fibre breaks up into a brush of fine fibrils, which are said to terminate in rounded ends.

Figure 2 points to a multipolar (motor) nerve cell in the grey matter of the anterior horn of the spinal cord (AH sp). From this cell an unbranched axis-cylinder process (acp) passes, through the anterior nerve root (anr), peripheral motor nerve (pmn), down to the muscle (M) to which the figure 3 points.

The branched fibres of the multipolar nerve cell in the spinal cord terminate in a brush of fine fibrils, which are supposed to terminate in rounded or bulbous ends, and which are in close apposition with the terminal fibrils of the fibres of the pyramidal tract.

tail. The degeneration involves the whole of the lower segment of the neuro-motor nerve apparatus (see fig 26) including the muscle. When the nerve cell is destroyed by the inflammatory process, its axis-cylinder process, the motor nerve fibre which is

a direct continuation of that axis-cylinder process, and the muscular fibre with which that motor fibre is connected, not only degenerate, but are permanently destroyed; they atrophy and they remain permanently atrophied, for so far as we know, a nerve cell which is once completely destroyed is never restored.

Remember, then, that when the multipolar nerve cells of the anterior horn of the spinal cord are completely destroyed by an acute inflammation, or indeed by any other pathological change, permanent paralysis and atrophy are produced in the muscular fibres with which they are connected, which they innervate and nourish. The destruction of the nerve cell must be complete; it is probable that if the nucleus remains regeneration of the nerve cell may take place. The effects which follow acute destruction of the multipolar nerve cells in the anterior horn of the spinal cord are diagrammatically represented in fig. 26.

You are now in a position to understand the fact which I have already mentioned, viz., that in severe cases of poliomyelitis anterior acuta the anterior nerve roots which pass out from the affected region of the cord look grey, are shrunken and atrophied, and that the motor nerve fibres of the peripheral nerves which are continuous with these anterior nerve roots are in the same (atrophied) state. The degeneration in the peripheral nerves may not be visible to the naked eye, but it can be seen with the microscope. A peripheral (spinal) nerve contains sensory as well as motor fibres, and its motor fibres are usually derived from several roots. Consequently, although it may appear normal to the naked eye, it is found to contain atrophied and degenerated fibres when examined with the microscope.

Further, in severe cases of poliomyelitis anterior acuta, the muscles after death are found to be in a condition of atrophy and cirrhosis.

The paralysis which results from destruction of the multipolar nerve cells is attended with flaccidity and atrophy of the muscular tissue; hence the term acute *atrophic* spinal paralysis which has been applied to the disease.

The degeneration and atrophy in the peripheral nerves and muscles is attended with certain electrical alterations which are of great importance (see page 63).

The next point to which I wish to direct your attention is

this, that the inflammation chiefly affects those parts of the spinal cord in which the grey matter is most abundant, viz., the lumbar and cervical enlargements. Hence the paralysis chiefly involves the muscles of the lower and upper extremities—the limb muscles. But the lumbar and cervical enlargements are composed of several segments, and it comparatively rarely happens that all of the grey matter of the anterior horns in all of these segments is involved; in most cases, it is only the grey matter in some of the segments which is affected. In many cases, the inflammation is, practically speaking, limited to the lumbar enlargement, or rather to some of the segments of the lumbar enlargement. In others, but this is less common, it is limited to the cervical enlargement or to some of the segments of the cervical enlargement. In many cases, some of the segments of the lumbar or some of the segments of the cervical enlargements only are affected. As a rule, the inflammation involves the two symmetrical lateral halves of the segment or segments which are affected; but this is not always so. Sometimes it is only one horn in one or more segments which is implicated, at least in any marked and appreciable degree. Even when the inflammation is bilateral, i.e. when it involves the two halves of any given segment, it is by no means always (in fact very rarely) strictly symmetrical; for the intensity of the inflammation, in the two lateral halves of the segment which is affected, may be very different.

I shall return to this point when I come to speak of the distribution of the paralysis.

From these statements, it will be apparent to you that the extent and severity of the lesion, the number of segments affected, and the manner in which the affected segments are implicated are very variable. These variations are of great clinical importance; they are difficult to remember as mere pathological facts, but you will have no difficulty in appreciating their significance and in remembering the variations which different cases present, when you come to study the subject practically at the bedside. This is only another illustration of the fact that the clinical and systematic study of disease should go hand in hand. I have already more than once insisted upon this point. It is impossible to reiterate it too frequently. It is a disadvantage of modern medical education that the study of anatomy, physiology and

pathology is to some extent divorced from the study of clinical medicine and clinical surgery, and that the teachers of anatomy, physiology and pathology are not clinical physicians and clinical surgeons. But the disadvantage is unavoidable; the more purely scientific and the more purely practical (the clinical) parts of medicine are now both so vast that they can only be satisfactorily studied and taught by specialists. In consequence of this separation and differentiation, the student often fails to realise, when he is pursuing his anatomical, physiological and pathological studies, the facts which are of primary and chief importance for practical (clinical) purposes. Indeed, you will, I expect, find it necessary to return again and again to the study of anatomy, physiology and pathology now that you have commenced *medicine* and clinical medicine. You need not be surprised at this. Even the most experienced of us have to do so every day. It is only when one reaches the clinical side of the subject that one realises *the* points in anatomy, physiology and pathology which are of most importance. I have more than once met with students who seem to think that after they have passed an examination in anatomy, physiology and pathology, they have done with these subjects. It is perhaps difficult to credit this statement, but it is a fact. Instead of being done with these subjects, the student is then only in a position to make use of them. Although the practitioner may afford to forget his botany and natural history, he cannot afford to forget his anatomy, physiology and pathology. These subjects are the very fundamentals, the only true bases of intelligent diagnosis and practice. The more anatomy, physiology and pathology the practitioner knows, the better. There are some points, of course—some parts of these subjects—which as yet have little or no apparent significance or importance for the purposes of practice; but every fact, *provided only that it is a fact*, may at some time or another be found to have some direct bearing on practice and treatment; you can never tell when it may not be useful.

The pathological facts, then, which it is essential that you should remember in connection with the disease which we are at present considering (poliomyelitis anterior acuta) are:—(1) That the lesion is an acute inflammation of the anterior horn of grey matter; (2) That, in most cases, the inflammation is practically confined to this part of the cord; (3) That it chiefly or

exclusively involves the grey matter of the lumbar and cervical enlargements (the limb areas); that all the segments in these areas may be involved, but that in the great majority of instances some of the segments only are implicated; that the lesion is as a rule bilateral, but by no means always or usually symmetrical: that sometimes it is unilateral, one horn only being implicated: that the lumbar enlargement is much more frequently affected than the cervical, but that it is by no means uncommon to find the lesion affecting the cervical portion of the cord, perhaps only one-half of some of the cervical segments, alone; (4) That the severity of the inflammation is very variable; that in some cases it is slight, and the chief result is a temporary inflammatory œdema, but that in other cases (and this is much more frequent) it is severe, and is attended with more or less destruction of the inflamed tissues; (5) That the effect of the inflammation is to temporarily arrest or permanently destroy the function of the active nerve elements (the cells and nerve fibres) of the anterior horn, paralysis (temporary or permanent) being the result; (6) That a nerve cell which is once completely destroyed is, so far as we know, never restored; and (7) That the result of destruction of a nerve cell in the anterior horn is degeneration and atrophy of its axis cylinder process, of the motor nerve fibre which is a direct continuation of that process, of the motor nerve-endings in which that fibre terminates, and of the muscular fibres with which those motor nerve-endings are connected.

I hope I have succeeded in making these points clear to you, for unless you thoroughly understand them you will find it difficult to understand the clinical phenomena of the disease. Further, you must remember that many of the points to which I have alluded (I refer more particularly to the conditions which result from destruction of the multipolar nerve cells) afford a key, not only to the symptomatology of poliomyelitis anterior acuta, but to the symptomatology of many of the other diseases of the nervous system which we shall afterwards have to consider. Some of the physiological and pathological questions which I have brought before your notice in this lecture are of the very first importance. I shall constantly have to refer to them in future lectures. We are continually making use of them at the bedside. It is for these reasons that I have con-

sidered them in such detail. I would urgently advise you therefore to lose no time in carefully revising the notes of this lecture, and in impressing the chief facts which I have detailed to you upon your memories. It is essential that you should make these facts part and parcel of yourselves, and that you should get such a knowledge of them that you can never forget them; that you should think them over, thoroughly digest them, assimilate them, master them, *know* them, so that you do not require to make an effort of memory to recall them. I assure you that the slight labour involved will be well spent. You will find that many of the facts which I have brought before your notice in this lecture will be of every-day use in practice; they will help you to unravel the symptoms in many obscure cases of nerve disease; they will aid you in diagnosis; they will give you important information for the purposes of prognosis; and they will be useful to you in respect to treatment.

LECTURE IV

POLIOMYELITIS ANTERIOR ACUTA (*Continued*)

Etiology.—Let us now turn to the etiology of the disease. Poliomyelitis anterior acuta is essentially a disease of childhood ; but it occasionally occurs both in older children and in adults ; the great majority of cases occur between the end of the first and the end of the third or fourth year. The disease is comparatively seldom met with before the age of six months ; it is very rare in middle life, and is practically unknown in old age. Many of the cases which were formerly described as poliomyelitis anterior acuta in the adult were probably cases of multiple neuritis or of ordinary myelitis which happened to be accidentally limited to the anterior horn of grey matter. It is of course quite impossible to distinguish during life the system disease poliomyelitis anterior acuta and an ordinary myelitis limited to the anterior horn of grey matter. The symptoms in the two cases are absolutely the same. Nevertheless there are some reasons for believing that the inflammation in the two conditions is due to a different cause. I shall return to this point presently.

Poliomyelitis anterior acuta is rarely hereditary ; a few cases have been reported in which several members of the same family, in the same or different generations, have been affected. I have myself met with some cases of poliomyelitis anterior acuta in which there seemed to be a distinct hereditary influence ; for example, three children in a family were affected with Friedreich's ataxia, and the fourth had well marked infantile paralysis ; an uncle of these children had also suffered from poliomyelitis anterior acuta.

The subjects of the disease sometimes come of a nervous stock. The hereditary influence is then apt to manifest itself indirectly, just as it does in so many other diseases of the nervous system. What I mean to say is this, that in many

cases in which a neurotic inheritance is handed on from parent to child, the same form of disease is not manifested in succeeding generations. For example, one of the parents may be nervous, neurasthenic, or 'peculiar,' the daughters hysterical, the sons drunkards, the grandchildren epileptic or insane, and so on. It may, I think, be doubted whether, in many of the cases in which a child of neurotic parents is attacked by poliomyelitis anterior acuta, the hereditary influence is a point of much importance; it is probably in many cases a mere coincidence.

The previous state of health of the patient is not an etiological factor of much importance. The disease often occurs in robust and healthy children, who were in perfect health up to the commencement of the attack. In some cases, the disease develops during teething; but it is doubtful if the irritation of teething has much to do with the production of the disease, for in the majority of cases teething cannot be blamed. Occasionally the disease follows, and seems to be due to, exposure to cold. One of the most important facts connected with the etiology of the disease is the circumstance that cases occur much more frequently in summer than in winter. This is of course only a general statement; for the disease is frequently developed during winter and cold weather. The explanation of this seasonal fact is not yet clear. Occasionally poliomyelitis anterior acuta follows or accompanies an acute febrile disease, such as scarlet fever; but this is not common. Cases of this kind, supposed poliomyelitis anterior acuta developing after measles or scarlet fever, are usually cases of peripheral neuritis. About a year ago, I saw with Dr. Murdoch Brown a case which seemed to be a typical example of poliomyelitis anterior acuta, in a well-grown girl aged 12; the disease had developed during convalescence from a severe attack of measles. The lower extremities were paralysed; the muscles were flaccid and markedly atrophied; the faradic contractility was abolished; the knee-jerks could not be elicited. Sensation of all kinds in the lower extremities was absolutely unimpaired. There was no affection of the bladder or rectum. The paralysis had developed acutely. In short, the case seemed to be typical in every respect; and as the paralysis was very extensive, the muscles markedly atrophied and the faradic

irritability lost, a very bad prognosis was given. But the prognosis was wrong. The patient completely recovered. The case was clearly one of peripheral neuritis, not of poliomyelitis anterior acuta; the recovery conclusively proved this; such a severe poliomyelitis anterior acuta would not have been completely recovered from. The peculiarity of the case—and this was the cause of the error in diagnosis—was the complete absence of sensory symptoms. I shall return to this point again when I come to speak of the diagnosis and prognosis.

Traumatic injury seems occasionally to be a predisposing or exciting cause; but it probably is a rare cause. The parents, it is true, often think that the disease is due to an injury or fall, and they sometimes very unjustly blame the nurse for having caused it; but in many cases of this kind, the supposed injury or fall had probably nothing to do with the production of the disease. It is only when the disease immediately follows a distinct injury, that a suspicion of traumatic influence should be entertained. Previous ill health, in fact anything which causes depression, may probably act as a predisposing cause; but as I have already told you, in many cases the subjects of poliomyelitis anterior acuta are, up to the time of the attack, unusually robust and strong.

The age at which the majority of cases occur (between the end of the first and the end of the third or fourth year) suggests that the active development of the grey matter and the active process of training, so to speak, that is going on in the nerve cells of the spinal cord, may have something to do with the development of the disease. During the second and third years of life, the child is learning, and perfecting itself in, the use of its limbs; muscular development is rapidly progressing, and a strain is consequently being thrown upon the multipolar nerve cells of the anterior horn; the great frequency of the disease during childhood is perhaps partly due to these conditions.

The following facts are, I think, highly suggestive of the view that poliomyelitis anterior acuta is a febrile disease, due to a definite toxic cause, rather than a mere local inflammation, viz. :— (1) the age at which the disease occurs; (2) the extreme rarity of second attacks; (3) the greater frequency of the disease during hot than during cold weather; (4) the occasional occurrence of 'runs of cases'; epidemics of poliomyelitis anterior

acuta have indeed been described, but, so far as I know, the disease is never infectious or contagious; (5) the fact that the fever and constitutional symptoms, which usher in the attack, are in many cases more marked than one would expect from the severity of the local lesion.

Now, these facts, when taken in combination, certainly seem to suggest that the inflammation in the cord and the fever and constitutional disturbance which are associated with it are due to a toxic cause. At present this is merely a theory; we have no definite evidence of a toxic cause; in particular, so far as I know, no organisms have been discovered in the inflamed tissue. But it must be remembered that the opportunity of examining the cord in the early stages of the disease very rarely occurs. It is highly probable, I think, that future observations will demonstrate the presence of some specific irritant, perhaps a micro-organism or the products of a micro-organism.

Most of you will before long be actively engaged in general practice. I would urgently impress upon you the importance of obtaining a post-mortem, if you should happen to meet with any case in which a child who is suffering from poliomyelitis anterior acuta dies (I do not say from the poliomyelitis, for the disease is very rarely of itself fatal) during the earlier (acute) stages of the disease. I will be much obliged if any of you will send me specimens of this kind. The affected portion of the cord should be placed (suspended by a thread attached to the membranes or nerve roots) in a wide-necked bottle filled with methylated spirit, and *immediately* despatched to me by parcel post.

Clinical History.—Let us now turn to the clinical history. In some cases, the attack is ushered in, or preceded by, premonitory symptoms, such as malaise, headache, irritability of temper, the child being obviously out of sorts, perhaps for a few hours or a day or two, before the characteristic symptoms develop. In the majority of cases, the onset is more or less abrupt, and is usually attended with a rapid rise of temperature, with fever in short, and with marked constitutional disturbance. The skin is generally hot and the pulse quick. In many cases, there is considerable prostration and restlessness. Anorexia and thirst are usually present; headache is common;

vomiting, muscular twitchings, an epileptic convulsion and slight delirium—symptoms which are apt to attend any sudden elevation of temperature in the child—are not uncommon. Exceptionally, especially in severe cases in young children, head symptoms of a more marked kind, such as decided delirium, repeated convulsions, and coma, occur.

After these symptoms—which are often thought to be of no importance, due, for example, to some derangement of the stomach, to teething, or to the commencement of some febrile disease such as scarlet fever—have lasted for some hours or perhaps a day or two, the characteristic paralytic symptoms are developed. The mother or the nurse notices that one of the limbs is flaccid and helpless. In some cases, but this is not common, the paralysis appears to usher in the attack, and to be followed, instead of preceded, by the febrile symptoms. It not unfrequently happens that the initial fever is so slight that it is not noticed. In these cases, the inability to move a limb or limbs may be the first thing which attracts the attention of the nurse or parent. It often happens, especially in very young children, that the loss of motor power is not noticed until the febrile symptoms subside, it may be several days after the paralysis has actually been in existence. It is very easy to understand how, in a young child, the fever and constitutional disturbance may occupy the chief place in the clinical picture and attract all the attention. The paralysis is often first noticed in the morning; the child goes to bed feverish and out of sorts, the next morning it is found to be paralysed.

The degree of fever and the severity of the constitutional symptoms are very variable; as a rule, the younger the child the greater the constitutional disturbance. Although, in the great majority of cases, some fever is present at the onset, the pyrexia is usually moderate in degree ($101\cdot5^{\circ}$ to $102\cdot5^{\circ}$). In some cases the fever is high (104° or even 105°). In others, the febrile disturbance is less than $101\cdot5^{\circ}$; these are the cases in which the constitutional disturbance is apt to escape attention, and in which the physician is apt to suppose that the paralysis has been developed without fever and other premonitory symptoms.

The paralysis.—In the great majority of cases, the paralysis is developed rapidly, in the course of a few hours; though, as I have already said, it is frequently not noticed until the febrile

disturbance comes to an end. In most cases—and this is a point of very great practical importance, for it is always a very satisfactory thing in such a disease as this to be able to afford any comfort to the parents—the height of the paralysis is rapidly reached. In the great majority of cases, the paralysis has already reached its greatest degree of development when it is first noticed, and any subsequent changes which occur are (usually) in the direction of improvement. In most cases, you can, therefore, with confidence assure the parents that the worst of the damage is already done when the paralysis is first discovered ; it is quite exceptional to find the paralysis extending, in any marked degree, after it is first noticed, although this does sometimes occur.

After what I have already said with regard to the pathology and morbid anatomy of the disease, you will easily understand that the extent, distribution and severity of the paralysis vary very greatly in different cases. The lower limbs are more frequently affected than the upper, but the upper limbs are often involved ; in rare instances all four limbs are implicated. Both lower extremities are frequently affected, both upper extremities are rarely involved in a marked degree. In a considerable number of cases, one limb only is affected. The latter statement applies more especially to a marked degree of paralysis ; it is in my experience very common to have one lower limb markedly, and the other only slightly, paralysed at the commencement of the case, and to find that in the course of time the limb which was severely affected remains more or less permanently affected, while the slightly affected one completely recovers. If this fact is kept in view, the monoplegic distribution of the lesion and of the paralysis will, I believe, be found to be much less common than some writers have supposed. You will see then that the different combinations which the distribution of the paralysis may present are very variable.

In some cases, the paralysis is not exclusively confined to muscles which are connected with the lumbar and cervical enlargements of the cord. The muscles of the back and neck are occasionally affected. In quite exceptional cases, the thoracic and abdominal muscles are involved. The diaphragm is said never to be affected. Muscles supplied by motor cranial nerves are very rarely paralysed. I have seen one or two cases in which

there was good reason to suppose that the facial muscles were paralysed as the result of poliomyelitis anterior acuta, and a few other cases of a similar kind have been described. It is said that the muscles of the head, eyeballs, pharynx, and larynx always escape. I have certainly not met with any case in which they were affected.

In most cases, the paralysis is bilateral, though not strictly symmetrical; the most common combination is to have both lower extremities affected, one more so than the other, and neither of them completely paralysed, at all events after the first effects of the lesion have passed away. In short, the paralysis rarely involves all the muscles of the affected limb or limbs. But cases do occasionally occur in which all the muscles of both lower extremities are completely and permanently paralysed, and in which the patient remains throughout life absolutely crippled, so far as his lower extremities are concerned. In a case which I have at present under observation (see Fig. 25), the lower extremities are completely paralysed; they hang like useless appendages, now some months after the onset of the disease. But in most cases, it is only some of the muscles of the limbs which are involved. Further, I would particularly ask you to remember that in many cases the whole of the affected muscles are not implicated; some of the muscular fibres in a given muscle may be paralysed, some may escape. This is a most important fact for prognosis and treatment.

As a rule, the paralysis affects muscles which are in the habit of functioning together; it is, in short, distributed physiologically rather than anatomically. The explanation of this fact is to be found in the circumstance that the nerve cells are grouped in a definite physiological order in the anterior cornua and segments of the spinal cord, so as to permit of the co-ordination of motor impulses. Hence, muscles which lie in anatomical juxtaposition in a limb, but which are innervated from different levels of the spinal cord, are not always affected. In some cases, for example, in which the muscles of the forearm are paralysed, the supinator longus escapes; this is sometimes termed the *lower arm type* of the disease; while, in other cases, the supinator longus (a muscle of the forearm) is affected, together with the biceps, brachialis anticus and the deltoid (muscles of the upper arm); this is termed the *upper arm type* of the disease.

The paralysis, then, is usually arranged in a physiological rather than an anatomical form. There are two reasons why

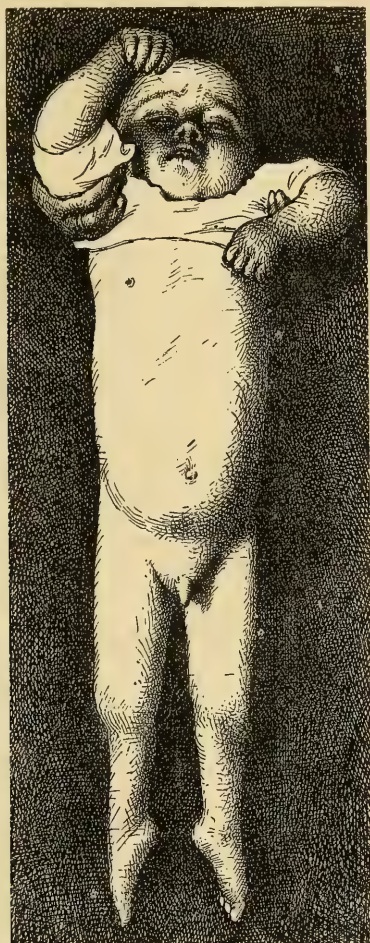


FIG. 25.—*Severe poliomyelitis anterior acuta ; the lower extremities are completely paralysed ; they hang like useless appendages ; the feet are everted.*

The drawing is an exact copy of an instantaneous photograph which was taken while the child was struggling.

part of a muscle, or some only of its muscular fibres may be permanently paralysed, viz. :—

(1) That many muscles receive their motor nerve supply

from more than one segment of the spinal cord. Consequently, if one segment only is affected, the muscular fibres of the affected muscle, which are supplied by unaffected segments, will escape. The nerve nucleus for the supinator longus muscle is situated in the upper part of the cervical enlargement (the segment from which the 5th root emerges), from which the upper arm muscles (deltoid, biceps and brachialis anticus) are innervated.

(2) That some of the nerve cells, in the affected segment or segments of the cord, may escape, while others are destroyed.

It is very important to remember not only that the maximum height of the paralysis is quickly reached, but that, in most cases at all events, a very considerable degree of improvement subsequently takes place; hence the term, *regressive* paralysis which has been applied to the disease. In speaking of the pathology, I emphasised the fact that some of the initial paralysis is usually due to a temporary arrest of function, the result of shock and the pressure which the inflammatory exudation causes. Now, the paralysis due to this cause may reasonably be expected to disappear as soon as the inflammatory exudation is absorbed, and the effects of shock and pressure are removed. While the paralysis which is due to destruction of the nerve elements (the motor nerve cells of the anterior horn) will remain; for, so far as we know, motor nerve cells which are once completely destroyed are never restored; nerve fibres are, however, often restored. This is a very important fact, for it explains the remarkable difference in regard to the recovery of the atrophied and paralysed muscles which exists between poliomyelitis anterior acuta—a lesion of nerve cells—and peripheral neuritis (alcoholic paralysis, for instance)—a lesion of nerve fibres. The extent and degree of the damage to the nerve cells is *the* point which determines the amount of the permanent paralysis which will remain.

Remember, then, that in the slight cases the paralysis may in great part disappear when the inflammation subsides; and that, as a matter of fact, in almost every case, some of the paralysis is recovered from. It is only in exceptionally severe cases that the initial paralysis is equal to the ultimate (permanent) paralysis; in other words, it is only in exceptional cases, that all of the nerve cells connected with the muscles

which are paralysed at the commencement of the case are permanently destroyed.

The condition of the reflexes.—The next point to which I must direct your attention is the condition of the reflexes. In severe cases, all of the motor impulses (voluntary and reflex) which pass through the inflamed portions of the anterior

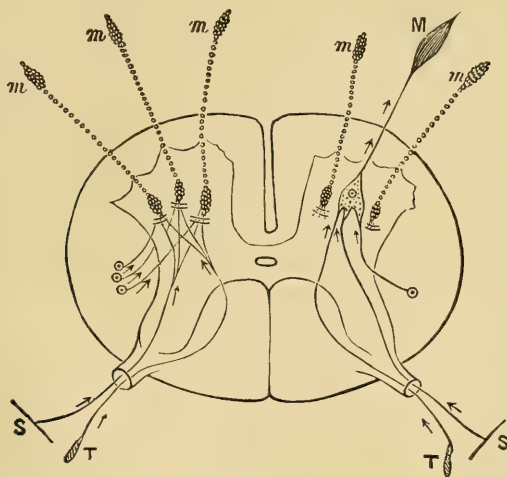


FIG. 26.—Diagrammatic representation of the symptoms which result from acute destruction of the anterior cornua of the spinal cord.

On the left side the destruction of the nerve cells is complete; the anterior nerve roots, motor nerve fibres, and muscles, which they supply, are all degenerated; there is a total 'block' to the passage of voluntary-motor and reflex-motor impulses. On the right side, two-thirds of the motor cells are destroyed; two-thirds of the muscular area connected with the right anterior cornu are degenerated and atrophied; one-third (M) remains healthy, and can be made to contract by voluntary or reflex-motor impulses.

horns are necessarily interrupted during the earlier periods of the case. This statement is of course not synonymous with the statement that in infantile paralysis the reflex movements are abolished. When, for example, the cervical enlargement is implicated, there may be complete paralysis and loss of reflex action in the upper extremity, without any paralysis or disturbance of reflex motion in the lower extremities. When, on the other hand, the lesion involves the lumbar enlargement, the reflexes of the lower extremity which pass through

the affected area of grey matter will be interfered with. In some cases, the interference with reflex action is only temporary; in others it is permanent. When, for example, the quadriceps extensor femoris is paralysed, the knee-jerk is abolished, and, if the quadriceps remain atrophied and paralysed, the knee-jerk remains permanently abolished. The condition of the paralysed muscles, in respect to the amount of muscular atrophy and the state of reflex action, will perhaps be better understood by reference to fig. 26.

The condition of the sensory functions.—Let us now turn to the condition of the sensory functions. We have seen that in the great majority of cases of poliomyelitis anterior acuta, the lesion is entirely, or for practical purposes entirely, confined to the anterior horn of grey matter—a motor area. Consequently the sensory functions of the cord and of the paralysed limbs are not interfered with. But in some cases, aching pains of a rheumatic-like (myalgic) character are complained of in the muscles or joints of the affected limbs; in some cases, there is tenderness on pressure over the affected muscles; and occasionally (but this is uncommon) tenderness on pressure may be elicited in the affected (degenerated) nerves. These myalgic, articular, and nerve pains are more common in adults and older children than in young children. In some cases, they are no doubt due to inflammation of the peripheral nerves, a neuritis which is associated with the cord lesion; but in other cases, they are, I think, probably due to the degenerative process which is going on in the motor nerve endings and the muscles. In this connection Dr. Sherrington's observations, which show that the muscular fibres are richly supplied with sensory nerves, are very important. Marked tenderness over the large nerve tracts is certainly suggestive of neuritis.

Any considerable degree of impairment of the sensibility of the skin is exceedingly rare. It is not uncommon to find some slight disturbances of sensation during the earlier stages of severe cases, but marked and persisting anæsthesia is quite exceptional. The presence of definite and distinct anæsthesia shows that the lesion is not confined to the anterior horn of grey matter. When sensory symptoms are well marked, the probability of the case being one of peripheral neuritis rather than of poliomyelitis should always be kept in view. The distinction is all-important

for prognosis. Perhaps in some of the cases in which there is marked anæsthesia the inflammation is not the result of the system disease poliomyelitis anterior acuta, but of an indiscriminate form of myelitis. It is generally supposed that ordinary myelitis very rarely (if ever) occurs in the child. I doubt whether this conclusion is altogether warranted. While I admit that the ordinary form of myelitis is very rare in children, I see no reason why it may not occur; just as the special form of myelitis which we are now considering—the system disease poliomyelitis anterior acuta—may occur in the adult.

The condition of the bladder and rectum.—In the great majority of cases of poliomyelitis anterior acuta, the functions of the bladder and rectum are not interfered with. Further, bed-sores are never developed. This is another distinguishing feature between the ordinary myelitis of adults, and this special (localised) form of myelitis. It is not uncommon, especially in severe cases in which the lumbar enlargement is affected, to have the functions of the bladder temporarily interrupted (for a few hours or days) at the commencement of the attack, as the result of shock and inhibition. But this is nothing more than we would expect. In typical cases of poliomyelitis anterior acuta, persistent and permanent paralysis of the bladder or rectum are practically speaking unknown. Marked paralysis of the bladder and rectum and bedsores are so rarely the result of the system disease poliomyelitis anterior acuta, that in any case in which they did occur in the child I should strongly suspect that the myelitis was indiscriminate in character.

The condition of the paralysed muscles.—Let us now turn to the condition of the paralysed muscles, a point of very great importance.

We have seen that the paralysis rapidly reaches its maximum degree and extent, and that although some of the initial paralysis usually disappears when the inflammation subsides, in well-marked cases, certainly in all severe cases, some paralysis, some weakness in the muscles usually remains.

The paralysis which results from poliomyelitis anterior acuta is essentially a flaccid paralysis. The paralysed limbs are limp and soft; this flaccid condition continues throughout the whole course of the disease. It is true that in long-standing (old) cases, the atrophied muscles may become rigid in conse-

quence of cirrhosis and cicatricial contraction; but this rigidity is not due to increased tonicity of the muscular fibres.

Now, it is of the utmost importance to draw a distinction between a flaccid paralysis and a spastic paralysis—a paralysis in which there is rigidity and usually no atrophy.

Paralysis of voluntary motion may of course be due to a lesion situated in any part of the motor nerve tract.

We divide the great motor tract, which extends from the motor centres in the cerebral cortex above to the muscles below, into two segments:—

(1) An upper segment which extends from the cortex of the brain above to the multipolar nerve cell in the spinal cord below, or rather to the terminations of the fibres of the pyramidal tract, for, as I have previously stated, it is now believed that the fine fibrils in which the fibres of the pyramidal tract terminate are not directly continuous with the branches of the multipolar nerve cells of the spinal cord; and

(2) A lower segment which extends from the multipolar nerve cell in the spinal cord above down to the muscle. (See fig. 24, page 42.)

The paralysis which results from a lesion (an organic lesion) in the upper division of the neuro-motor nerve apparatus is usually a 'rigid' paralysis: while the paralysis which results from a lesion in the lower division of the neuro-motor nerve apparatus is usually a flaccid paralysis. Further, the condition of the paralysed muscles in respect to their state of nutrition, their electrical reactions, and the condition of the reflexes, is markedly different in the two cases. I shall consider these points in more detail when I come to speak of spastic paraplegia. Suffice it now to say that the division of the cases of paralysis into the two groups (upper and lower types, as they may be termed) is of the greatest practical importance. The multipolar nerve cell, which is the trophic nerve centre for the lower segment, marks the point of division, for the dendritic branches of the multipolar nerve cell (as the branches other than the axis-cylinder process have been termed) and the fine network of nerve fibrils which they form should be regarded as essential parts of the multipolar cell, i.e. of the multipolar cell system, as it may be termed. The essence of the whole thing, so far as the condition of the paralysed muscles is concerned, is whether the func-

tional activity of this trophic nerve centre is retained or not, and whether that trophic influence is able to reach the muscle. This is the key to the whole position.

Now, in poliomyelitis anterior acuta, we find as a matter of fact that in the course of ten or fourteen days the paralysed muscles, or some of them, begin to waste. The atrophy rapidly progresses; and, in the course of six or eight weeks, the muscles which are seriously damaged (those muscles connected with multipolar nerve cells which are destroyed) may have lost half their bulk.

This rapid atrophy is associated with very definite histological changes both in the paralysed muscles and in the affected nerves. I repeat that these changes result from the destruction of the trophic centres—the multipolar nerve cells in the anterior horn of the spinal cord. In consequence of the withdrawal of the trophic nerve influence, the axis-cylinder process, the motor nerve fibre which is continuous with it, the motor nerve endings, and the muscular fibres which those motor nerve endings innervate (i.e. the muscular fibres connected with the cell which is destroyed) rapidly degenerate.

The histological changes are exactly similar to those which follow section of a motor nerve. The white sheath of the nerve is split up into minute globules, the axis-cylinder is broken into fragments, the nuclei of the nerve fibre become enlarged; in short, the structure and function of the nerve and of its motor nerve-endings are (for the time at least) entirely destroyed and abolished.

Well marked histological changes are at the same time developed in the paralysed muscles. The nuclei of the blood vessels, of the connective tissue and of the muscular fibres themselves, enlarge and proliferate; the transverse striæ disappear; and finally, if the atrophy and degeneration are permanent, the muscular fibres are replaced by fibrous tissue and fat. In short, in incurable cases a cirrhotic condition is produced.

Now, in the living patient it is difficult or impossible to detect the presence of muscular atrophy during the early stages of the case, for the degeneration and atrophy are not immediately established; time is required for their development. But in the course of a month or six weeks—I am referring to severe cases—the atrophy is very apparent. I have already stated that at

the end of six or eight weeks the muscles which are severely affected may have dwindled to half their normal size.

From these statements you will readily perceive that during the early stages of the case (the first two or three weeks) it is impossible from the mere appearance of the paralysed muscles to determine whether they are undergoing the degenerative process and whether they are likely to be permanently paralysed or not. But there is one condition which shows with much more certainty that this degenerative process is going on—I refer to the electrical condition of the affected muscles and nerves. When the paralysed muscles and nerves are undergoing the degenerative changes to which I have just referred, certain electrical changes which are grouped together under the term—*the reaction of degeneration*—are developed. The changes can be detected as soon as the degeneration in the nerves is well established—that is to say at the end of a week or ten days—at a time, you will observe, when the muscular atrophy is not yet apparent.

The reaction of degeneration is of great importance for the purposes of diagnosis, prognosis and treatment; and as this is the first occasion on which I have had to mention it, it will perhaps be well for me to describe it in detail. I shall have to refer to this important electrical change again and again throughout the course, and I want you once for all to understand exactly what it means.

The electrical changes which constitute the reaction of degeneration are a great puzzle to students, but they are not difficult, they are in fact exceedingly simple if you go about their study in the right way. There is of course nothing more difficult than to learn off by rote—and that is the usual way in which students try to learn them—the changes which constitute the reaction of degeneration. Studied in that way, the reaction of degeneration is one of those things ‘that no fellow can understand’; for to learn off by rote, in order to repeat like a parrot, these electrical alterations, a very considerable effort of memory is required; but if you study these changes intelligently they are easily understood and easily remembered.

LECTURE V

POLIOMYELITIS ANTERIOR ACUTA (*Continued*)

At the end of the last lecture, Gentlemen, I was about to describe and explain the electrical alterations which we term '*the reaction of degeneration.*'

You are aware that two forms of electricity (the interrupted and the constant forms of current) are chiefly employed in medicine. The effect of both forms of current (when sufficiently strong to produce any obvious, external effect), is to excite the functional activity of the muscle or motor nerve to which the electricity is applied. But the constant current only excites function (i.e. produces muscular contractions) when its strength is suddenly increased or suddenly diminished. So long as a constant current of electricity is flowing through a motor nerve or muscle, there is no muscular contraction, unless indeed the current is very strong; but I am speaking of currents of the ordinary strength which we use in medical practice, not of very strong currents. Remember, then, the constant current only produces muscular contractions when its strength is suddenly increased or diminished—in other words, when you make it or break it.

The interrupted current may be regarded as a continuous current which is being very rapidly made and broken; hence when an interrupted current (of sufficient strength) is applied to a motor nerve or muscle, a continuous contraction is produced; the muscular contractions follow one another with such rapidity that they fuse, as it were, into one. In short, a condition of tonic spasm is the result.

These are the effects which are obtained in health, and it matters not whether you apply the electricity to the nerve or to the muscle; the effect in both cases is the same. This is due to the fact—and it is a very important point—that in health it is

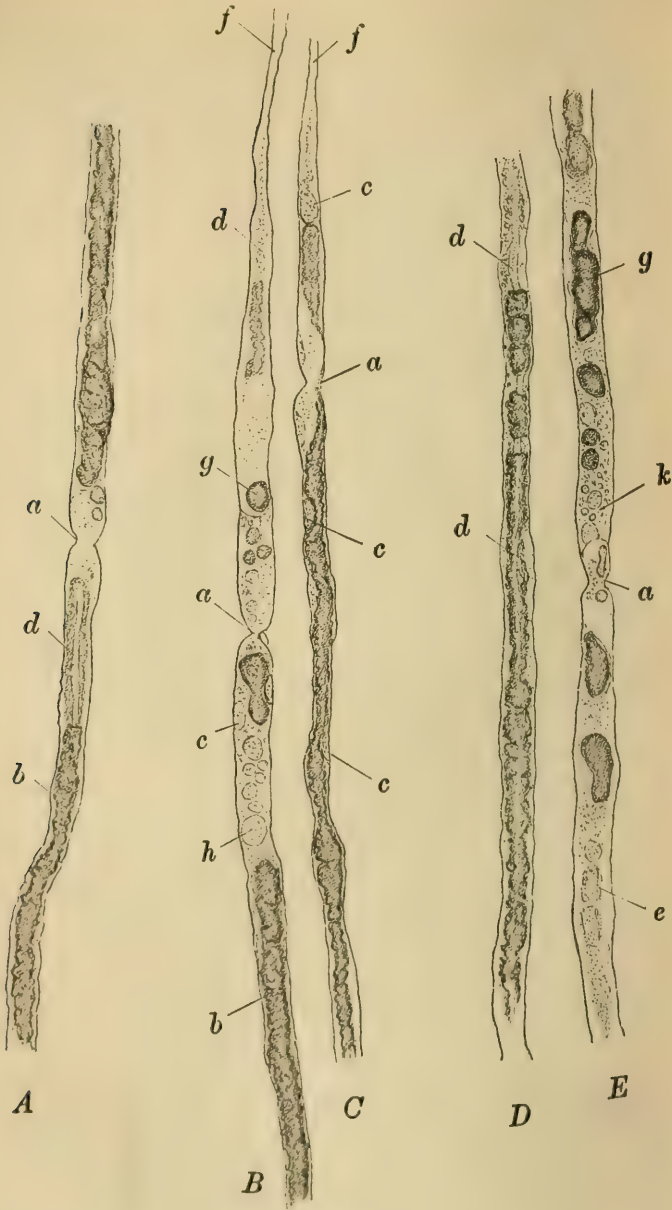


FIG. 27.

impossible to stimulate the muscular fibres without at the same time stimulating the motor nerve-endings which the muscle contains. This is one of the secrets, if I may use the term, of the reaction of degeneration. In fact, the contraction which results in health from the direct application of electricity to a muscle is due to stimulation of the motor nerve-endings rather than to stimulation of the muscular fibres themselves. We shall see the importance of this presently.

In health, the contractions which are produced by electrical stimulation are short, sharp, and lightning-like; a flash of muscular contraction, as it were, is produced, when the constant current is made or broken.

Now, in poliomyelitis anterior acuta, the axis-cylinder processes, the motor nerve fibres which are continuous with them, and the motor nerve-endings in the muscle are all degenerated; they are all broken up and destroyed. The histological changes in the degenerated nerves are similar to the changes which occur in the peripheral end of a motor nerve after section, and in the peripheral nerves in many forms of neuritis. (See fig. 27.)

Further, the muscular fibres themselves are undergoing an atrophic and degenerative change.

The effects of electricity applied to muscles and nerves which are degenerated in this way are, as you might naturally expect, quite different from those which are produced from the application of electricity to healthy muscles and nerves; hence the term '*reaction of degeneration*' which has been applied to these electrical

DESCRIPTION OF FIG. 27.

Fig. 27 is a camera lucida drawing of longitudinally divided nerve tubules (*A, B, C, D, and E*), from the posterior tibial nerve in a case of diabetes mellitus and perforating ulcer of the foot. Stained with osmic acid and picro-carmin, and mounted in Farrant's solution. Magnified, Hartnack; ocular 3; objective 9; immersion tube drawn out; and drawing reduced from 7 to 6½ inches.

The letters *a, a, a*, point to Ranvier's nodes; *b, b*, to the myeline sheath at points where it still remains entire (unsegmented); *c, c, c*, to nuclei, some of which are proliferating; *d, d, d*, to the axis-cylinder; *e*, to a greatly enlarged axis-cylinder in the process of breaking down; *f, f*, to empty and shrivelled portions of the nerve tubules; *g, g*, to portions of broken-down (segmented) myeline sheath; *h*, to a large globular mass, which appears to be a broken-down portion of the myeline sheath, but which does not stain (or is only very faintly stained) with osmic acid; *k*, to a mass of small globules and granules, also unstained with osmic acid, in the neighbourhood of a Ranvier's node. Many other globular masses, unstained by osmic acid, are seen in the neighbourhood of *h* and *k*.

alterations. I repeat that the electrical changes are the result of the degenerated condition of the nerve and muscle. This is the key to the position, the explanation of the whole thing. Whenever you think of the reaction of degeneration, you should picture to yourselves, you should call up in your mind's eye, these degenerative changes, which (in the case of poliomyelitis anterior acuta) extend, you will remember, from the multipolar nerve cells in the anterior cornu of the spinal cord right down to, and into, the affected muscles. If you remember these degenerative changes, you can predict, without any knowledge of the subject at all, what some of the electrical alterations will necessarily be. What effect would you expect electricity to produce in a nerve which is degenerated and destroyed? Why, of course, no effect at all. And I would beg of you to remember that the motor nerve-endings in the muscle are degenerated and destroyed.

This is the *first fact* in the reaction of degeneration; when the electricity (electrode) is applied to the (degenerated) nerves there is no response, either to the interrupted or the constant form of current.

We may represent this, the first fact in the reaction of degeneration, by the following formula:—

$$(1) N \cdot \frac{I.c.}{C.c.} = 0.$$

In other words, the nerve (N) (which is degenerated and destroyed) when stimulated, either by the interrupted (I.c) or the constant current (C.c.), does not respond, i.e. there is no muscular contraction, (=0).

The *second fact* in the reaction of degeneration is explained by two circumstances, viz.:—(1) That, when the motor nerve-endings are degenerated, the muscular (or sarcous) substance only remains to be stimulated; and (2) That the muscular (sarcous) substance does not contract to (i.e. its function is not excited by) an electric current of very brief duration.

Now, the interrupted current is a current of brief duration; it may be regarded as a constant current which is very rapidly made and very rapidly broken. When, then, the interrupted current is applied to a muscle, whose motor nerve-endings are degenerated and destroyed, there is no response. In health, you will remember, an interrupted current which is directly

applied to a muscle produces muscular contractions by exciting the functional activity of the motor nerve-endings, rather than by stimulating the muscular fibres themselves. In poliomyelitis anterior acuta, the degenerative process removes, as it were, the motor nerve-endings; it produces exactly the same effect as the poison woorara does. Hence there is no response (no contraction) when the interrupted current is applied directly to the muscle.

This, the second fact in the reaction of degeneration, may be represented by the formula:—

$$(2) \text{ M. I.c.} = 0.$$

In other words, the muscle (M), when stimulated by the interrupted current (I.c.) does not contract, ($=0$).

The *third fact* in the reaction of degeneration is this, that the constant current when applied directly to a muscle whose nerve-endings are degenerated, does (usually) produce contractions. This fact is explained by the circumstance that the muscular (sarcous) substance is stimulated, thrown into contraction, by a slowly interrupted continuous current. This, the third fact in the reaction of degeneration, may be represented by the formula:—

$$(3) \text{ M. C.c.} = \text{contractions.}$$

But, as one would naturally suppose, the contractions which are produced are different from those which are obtained in health; for, as I have already repeatedly stated, the contractions which are produced in health by direct stimulation of a muscle, are due to the stimulation of the motor nerve-endings, rather than of the sarcous substance.

The abnormal contractions which are obtained by the direct application of a constant current to a muscle whose nerve-endings are degenerated (and which are due to stimulation of the sarcous substance), differ in three particulars from the normal contractions which result from the direct application of a constant current to (direct stimulation of) a muscle (and which are chiefly due to stimulation of the motor nerve-endings). They are as follows:—

(a) In the first place, the contractions which are produced

by this weaker (constant) current resemble the contractions of involuntary rather than of voluntary muscle. Instead of being short, sharp and lightning-like in character, like the normal contractions, they begin more gradually, they persist longer and decline more slowly. The contraction curve resembles that of an involuntary (non-striped) rather than of a voluntary (striped) muscular fibre. This fact, which is of great practical importance, may be represented by the following formula and diagram (see figs. 28, 29, and 30):—

M. C.c.=(a) more gradual contraction curve.

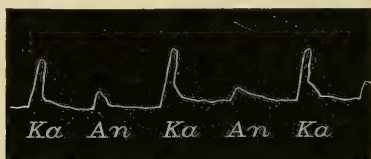


FIG. 28.—Curve taken from a healthy girl. 33 cells. KCC much more marked than ACC.—(After Erb.)

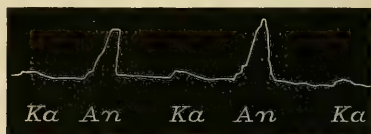


FIG. 29.—A case of chronic anterior poliomyelitis, with RD. Curve taken in the peroneal region. 33 cells. ACC notably greater than KCC.—(After Erb.)

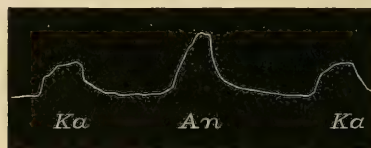


FIG. 30.—The same case. 40 cells in circuit. Ascendency of the ACC and sluggish character of the contractions well marked.—(After Erb.)

(b) In the second place, the muscle (usually) responds more readily than in health, a weaker current is (usually) sufficient to produce contractions. If you remember that the constant current is capable of exciting (stimulating) the sarcous substance, and that the degenerated sarcous substance is more easily

stimulated than the motor nerve-endings, you have an explanation of this fact in the reaction of degeneration. It may be represented by the formula:—

$$M. C.c. = (b) + \text{contractions.}$$

The increased excitability of the muscle to the galvanic current continues for a longer or shorter period in different cases. In incurable cases, after twenty or thirty weeks it gradually declines, and finally ceases (see fig. 33); then the muscle, like the nerve, ceases to respond to all forms of current. In cases which recover, the increased galvanic excitability may last for some weeks or even months, it then gradually diminishes, and the normal reactions reappear. (See figs. 31 and 32.) It is important to remember that in cases which slowly recover, the power of producing voluntary contractions in the paralysed muscles is usually regained before artificial contractions can be produced by applying either the continuous or the interrupted current to the motor nerve, in fact, in some cases it is impossible, even when the paralysis has to a large extent disappeared, to produce any contraction by means of electricity applied to the motor nerve trunk.

The following diagrams, taken from Erb, illustrate the course and duration of the 'reaction of degeneration' in different cases:—

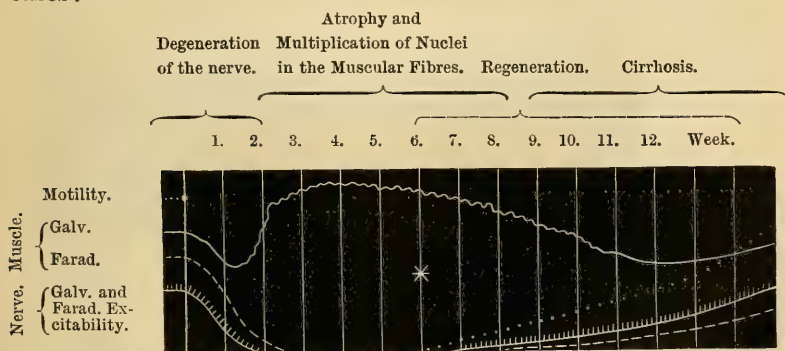


FIG. 31.—*The reaction of degeneration in a mild peripheral lesion (rapid recovery).*
—(After Erb.)

Motility, represented by the dotted line, ceases with the occurrence of the lesion; the galvanic and faradic excitability of the nerve gradually diminish, and at the end of two weeks disappear, the nerve trunk is now degenerated; the faradic excitability of the

muscle gradually diminishes, and ceases at the end of two weeks, the motor nerve-endings are now degenerated; the galvanic excitability diminishes for two weeks, it then usually increases, and the other phenomena of the 'reaction of degeneration' are present; the star (*) marks the return of voluntary motor power; the galvanic and faradic excitability of the paralysed nerve, and the faradic excitability of the paralysed muscle next reappear; the increased galvanic excitability of the muscle gradually diminishes, and the other phenomena of the 'reaction of degeneration' gradually disappear.

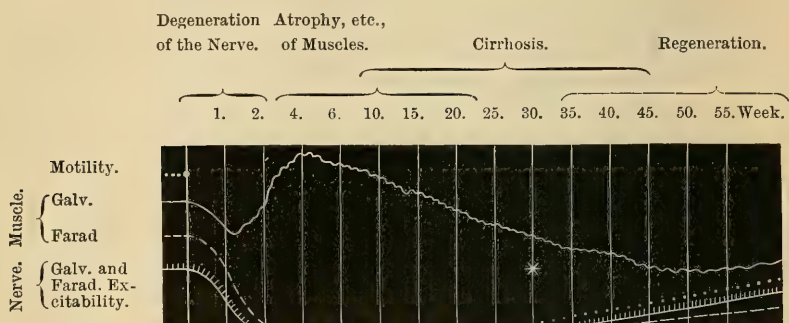


FIG. 32.—*The reaction of degeneration in a severe peripheral lesion (slow recovery).*
—(After Erb.)

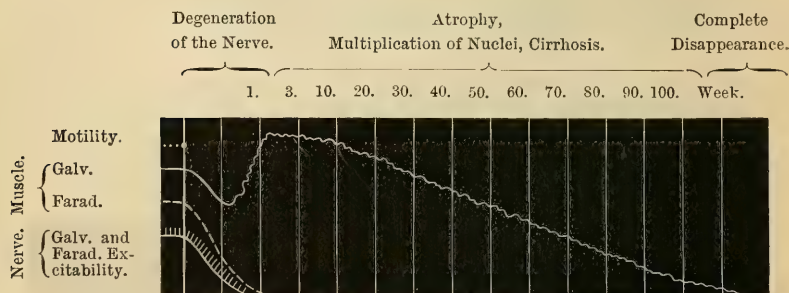


FIG. 33.—*The reaction of degeneration in a severe peripheral lesion in which there was no recovery.*—(After Erb.)

(c) In the third place, the order of the polar reaction, as we term it, is reversed, the anode taking the place of the kathode. This, which is the only difficult point (and fortunately it is by far the least important from a practical point of view), the only fact in connection with the reaction of degeneration which

demands any effort of memory, requires some explanation. All the other points which I have just mentioned are easily enough remembered, for they are based upon intelligible explanations and facts which are easily understood. But it is difficult to remember what the order of the abnormal polar reactions is unless you know and remember what the order of the normal polar reactions is.

Now, what is meant by the 'order of the polar reactions'? You are aware that there are two poles, the positive and the negative, the *anode* and the *kathode*, as they are technically termed. In using the interrupted current, there is no need to differentiate between these two poles, but it is a matter of great importance to differentiate between them when you are using the constant current for the purposes of diagnosis; because we find when we are using a weak constant current (1) that when we close (or make) the circuit, muscular contractions are more readily obtained than when we break (or open) it; and (2) that muscular contractions are more easily obtained when we close (or make) the circuit, with the negative pole or electrode (the *kathode*) on the muscle which we desire to stimulate, the anode being at the distant or neutral point, say, the nape of the neck or sternum, than when the positive pole is on the muscle and the negative on the neutral point. In other words, in health, *kathodal opening contraction* (K.c.c.) is greater than *anodal opening contraction* (A.c.c.). This is represented by the formula:—

$$\text{K.c.c.} > \text{A.c.c.}$$

It is not difficult to remember that the *kathodal closing contraction* is the contraction which is most easily (first) obtained in health, if, when you make the effort of memory, you picture yourself stimulating a muscle of the lower limb the neutral electrode being placed on the nape of the neck, and if you remember that the electric current is supposed to flow from the positive electrode to (*down to*) the *kathode* (*κατα*, down); in other words if you suppose that the electricity is flowing down from the nape of the neck to the leg, that is (*κατα*, down) from the anode on the nape, to the *kathode* on the muscle. I need not say this is merely an artificial guide to the memory.

I repeat that the 'order of the polar reactions' is the only

difficult thing to remember in connection with the reaction of degeneration; but even this is much less difficult to understand and remember than at first sight appears. I have given you an artificial 'tip' by which you may remember that in health the kathodal closing contraction is the first contraction to appear. Let me add that the corresponding kathodal opening contraction is usually the last to be developed.

I have already stated that when the reaction of degeneration is present, i.e. on stimulating the degenerated muscle directly by the constant current, the order of the polar reactions is reversed—the anodal closing contraction usually appears before (or at the same time as) the kathodal closing contraction. This may be represented by the formula:—

$$M.C.c. = A.c.c. > K.c.c. \text{ or } A.c.c. = K.c.c.$$

The fact that the anodal closing contraction usually appears before or at the same time as the kathodal closing contraction is practically all that it is necessary to remember with regard to the order of the polar reactions.

It may, however, be well to state that the normal and abnormal order of the polar reactions is more complicated than this statement would imply.

The full order of the normal polar reactions.—Since there are two poles (the *anode* or positive, and the *kathode* or negative), and since muscular contractions may occur on making or closing, and on breaking or opening the current, it follows that there are four possible forms of contractions, viz.:—

(A. When the negative pole is on the muscle or motor nerve, the positive pole on the distant or neutral point.)

1. The contraction which occurs on closing=the kathodal closing contraction K.C.C.

2. The contraction which occurs on breaking or opening=kathodal opening contraction K.O.C.

(B. When the positive pole is on the muscle or motor nerve, the negative pole on the distant point.)

3. The contraction which occurs on closing=anodal closing contraction A.C.C.

4. The contraction which occurs on opening=anodal opening contraction A.O.C.

These four forms of contraction usually occur in a definite order with currents of increasing intensity, viz. :—

1. K.C.C.
2. A.C.C.¹
3. A.O.C.²
4. K.O.C.

As the current strength increases so does the intensity of each contraction. We may, therefore, represent the character of the contractions with a gradually increasing strength of current as follows :—

1. *Very feeble* current. No contraction either on opening or closing with either pole.
2. *Weak* current, K.C.C.
3. *Somewhat stronger* current, K'.C'.C'. + A.C.C.
4. *Still stronger* current, K''C''C'' + A'.C'.C'. + A.O.C.
5. *Very strong* current, K'''C'''C''' (kathodal tetanus) + A''C''C'' + A'O'C'. + K.O.C.

In health, then, constant or galvanic currents of moderate strength only produce contractions when their strength is suddenly altered (i.e. when the current is made or broken); the contractions which result from the application of the current to the motor nerve or to the muscle itself, are identically the same, and appear in a definite 'polar' order, the kathodal closing contraction being the first, and the kathodal opening contraction the last to be manifested.

The full order of the abnormal polar reactions (when typically developed) *in the reaction of degeneration* is usually as follows :—

- | | | |
|--------------------------------------|----|-------------------------------------|
| 1. A.C.C. | | 1. K.C.C., A.C.C. |
| 2. A.C.C. + K.C.C. | or | 2. K.C.C., A.C.C. + A.O.C. |
| 3. A.C.C. + K.C.C. + K.O.C. | | 3. K.C.C., A.C.C. + A.O.C. + K.O.C. |
| 4. A.C.C. + K.C.C. + K.O.C. + A.O.C. | | |

I say the full order of the polar reactions, in those cases in which the reaction of degeneration is present, is *usually* that given above. But this statement must not be taken too

¹ The A.C.C. and A.O.C. sometimes appear simultaneously. Sometimes the A.O.C. appears before the A.C.C.

² Dr. de Watteville points out that the occurrence of AOC is determined not only by the strength of the current but also by the time the current has been flowing through the nerve before it is broken.

absolutely. It is often difficult to demonstrate these alterations in the living patient. In a considerable number of cases the partial or imperfect form of the reaction of degeneration which is represented in fig. 34 is alone developed.

Such are the electrical alterations which constitute the reaction of degeneration.

I have occupied some time in describing them, but I assure you it has not been misspent. I shall frequently have to mention the reaction of degeneration in this course of lectures, but I shall not explain it again. I hope you will endeavour to thoroughly understand and master it, so that whenever in future

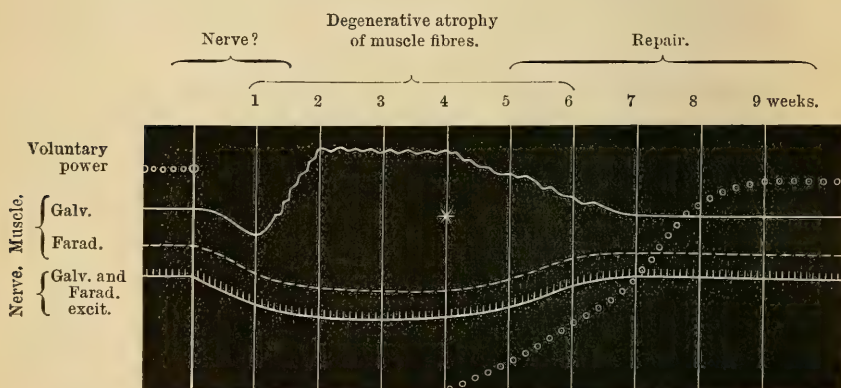


FIG. 34.—Graphic representation of the partial form of RD. The faradic and galvanic excitability of the nerve, and the faradic excitability of the muscle, are but slightly lowered. Voluntary power soon restored. Recovery rapid and complete. There was probably no degeneration of the nerve.—(After Erb.)

I mention the term 'reaction of degeneration,' you will know exactly what I mean. Further let me beg of you, not merely to be content with understanding it in the lecture-room, but let me urge you to try and work it out for yourselves at the bedside. Unless you do this, your knowledge will be merely theoretical, it will have no real practical value. A working knowledge of the art and method of applying electricity, both for the purposes of diagnosis and treatment, can only be acquired by much practice and experience. It can only be gained by patiently and laboriously observing the effects which the different forms of current produce, in health and disease, in the living man. You should commence by observing the effects in health. I would

advise you to practise on yourselves and your fellow-students; you may feel quite sure that you will not discover any serious nervous lesion; you can safely and comfortably therefore pursue the study. There is no risk in this. It is different when the student commences to study the heart sounds by applying the stethoscope (a binoral) to his own person or that of a fellow-student. By this procedure, murmurs or other supposed abnormalities in the heart sounds are often discovered by the beginner, who may in consequence be terrified by the notion that he is suffering from some grave cardiac disease. A session seldom passes without at least one case of this kind coming under my notice. I have almost always found the cardiac sounds beautifully and typically normal in these cases. I have never as yet met with a single case, where supposed murmurs have been discovered in this way, in which there was any organic cardiac disease.

Further, I am quite sure that you will not use too strong currents upon yourselves, whatever you may do upon your patients. It is advisable for this reason to begin the practical study of electricity by applying the electric current to your own bodies, and to feel and realise in your own persons the sensations which electric currents are capable of producing. Patients very much object to be stimulated in the way that they sometimes are by students and beginners.

Before leaving the subject, let me repeat that some of the electrical alterations which I have just described as characteristic of the reaction of degeneration in its typical and fully developed form, cannot always be elicited at the bedside. In not a few cases, the abnormal order of the polar reactions is not developed; in other cases, the increased excitability to the constant current is not very apparent. The facts which are of real practical importance are:—(1) That the irritability of the affected nerves is completely lost, when the lesion is severe and the nerve degeneration and destruction are complete; (2) That the faradic irritability of the paralysed muscles is completely lost; and (3) That the affected muscles, which are incapable of responding to the interrupted current, can (usually) be thrown into contraction by the galvanic current and that their contractions are slower (less sharp and lightning-like) than in health and more apt to become tetanic.

Now let us return to the clinical history of poliomyelitis anterior acuta. I repeat that those muscles which are seriously affected exhibit the 'reaction of degeneration' in a more or less typical form; and that the important electrical alterations, which constitute the reaction of degeneration, can usually be detected, in well-marked (severe) cases, at the end of a week or ten days after the onset of the attack. Let me again emphasise the facts that at this early stage of the paralysis, the muscles, though flaccid, are not distinctly atrophied; and that the electrical test is the only means of deciding, (but even it perhaps cannot always be absolutely relied upon) whether the motor nerve fibres are degenerated. In a case of poliomyelitis anterior acuta, the presence of the reaction of degeneration proves that the lesion in the cord is severe. Those muscles which present the reaction of degeneration, (provided that the degeneration in the nerve fibres is due to destruction of the nerve cells, but this is, in some cases, a difficult or impossible matter to determine) will probably remain more or less permanently paralysed and atrophied.

I have now described to you, and I hope enabled you to clearly realise and understand, the chief points connected with the symptomatology of this disease. We have seen that, in most cases, the disease is rapidly developed with fever and constitutional symptoms; that paralysis is quickly established; that the fever generally lasts for a few hours or days and then subsides, but that the paralysis remains; that the extent, distribution and severity of the paralysis are very variable in different cases, but that in most cases (in all but the very slight ones some muscles or parts of muscles are so severely affected that they present the reaction of degeneration in a more or less typical form and that they rapidly atrophy; that the reflex movements which pass through the affected area of the spinal cord are interrupted and abolished; and that there are, as a rule, no sensory phenomena, no derangements of the bladder or rectum, and no bed-sores.

Now, in order to complete the clinical history it only remains to describe the changes which subsequently take place in the paralysed muscles. We have seen that the greatest degree of the paralysis is quickly reached, and any changes which subsequently occur are in the direction of improvement. This, at all

events, is the general rule. But you must remember that time is required for this improvement. Except in the very mildest

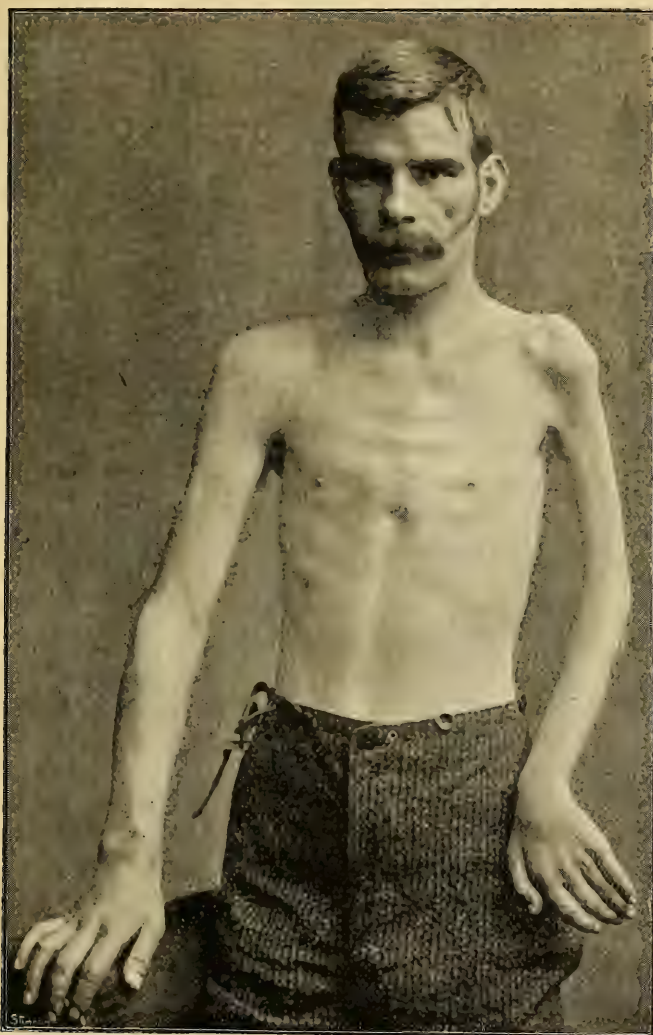


FIG. 35.—*Poliomyelitis anterior acuta*; paralysis and atrophy of the left arm; the muscles of the shoulder girdle and of the upper arm are more affected than those of the forearm and hand.

cases, there can be little or no improvement for some weeks at least; until, in fact, the pressure which the inflammatory œdema

and exudation cause, is removed. Further, the improvement steadily but slowly continues until the functional activity of the affected nerve elements (the nerve cells and fibres) is restored, or so far restored as the damage which they have sustained will in each case allow. This restoration requires time; it is rarely perfect; some permanent damage generally remains; in very severe cases, a whole limb or limbs may remain absolutely paralysed and permanently atrophied.

In the case from which this drawing was made (see fig. 35), the upper arm is permanently wasted and paralysed; the muscles of the forearm are, you will observe, less severely affected. In this case (see figs. 36 and 37), the left leg has been very seriously damaged; it is several inches shorter than its fellow on the opposite side.

When the affected muscles are well covered by subcutaneous fat, as is often the case in young subjects, the wasting may not be very apparent. In thin subjects, especially in muscular adults, and in old standing cases, the disproportion between the affected and unaffected limbs is very striking.

We have seen that those muscles which are permanently paralysed and atrophied ultimately become transformed into fibrous tissue. Now, the shortening which results from the cicatricial contraction, as it is termed, in the muscle, is apt to produce marked deformity. This is one of the results of the paralysis which it is important to bear in mind. Various forms of club-foot and other deformities are met with as the result of poliomyelitis anterior acuta; some of them are produced in this way. Further, deformity may also be produced by the healthy muscles which, unopposed in their action in consequence of the paralysis, are pulling upon a limb or joint. The weight of the limb and the want of support, too, which attends the atrophied condition of the muscles may produce deformities and looseness in the joints. In cases of old-standing infantile paralysis, the ankle is often very 'wobbly' and loose (see fig. 37); this has to be attended to in the treatment. Atrophy of the deltoid and scapular muscles and the relaxation of the ligaments produced by the want of muscular support and the weight of the paralysed upper limb may allow the head of the humerus to fall away from the glenoid cavity (see fig. 35). In such cases, dislocation of the shoulder joint is much more readily produced than in health.

But the most marked and important deformities which result from infantile paralysis are due to arrested development of the bones of the affected limb. The fact that the bones of limbs which are permanently paralysed and atrophied do not

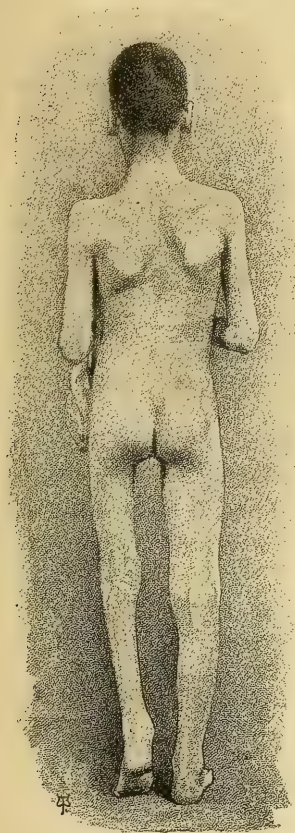


FIG. 36.

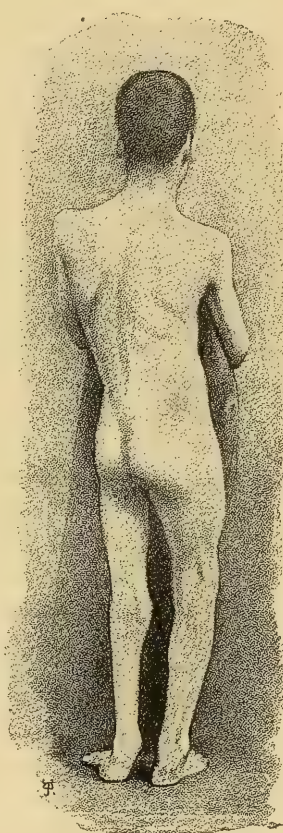


FIG. 37.

FIGS. 36 AND 37.—*Poliomyelitis anterior acuta* ; paralysis and atrophy of the left leg ; the muscles below the knee are more affected than those of the hip and thigh.

develop in the normal way seems to show that the bones like the muscles are nourished by the multipolar nerve cells in the anterior cornua of the spinal cord, or at all events that the trophic nerves for the bones pass through the anterior cornua. The arrested development cannot be merely the result of arrested

muscular development. This is proved by the fact that in pseudo-hypertrophic paralysis the bones grow and develop and attain their normal length (but not their normal thickness), notwithstanding the fact that the muscles which are attached to them are absolutely wasted and functionless. In a subsequent lecture I will describe a remarkable case in point.

In the upper extremity the non-development of the bones is of less importance than in the lower. Marked shortening of one lower extremity not only produces a limp, but it is apt to lead to secondary distortion of the spinal column; the whole body becomes twisted in consequence of the efforts which the patient makes to walk on the weak, short limb (see figs. 36 and 37). The muscular development in the sound limb, on which an excessive strain is necessarily thrown in standing and walking, is, in some cases, very great.

Limbs which are permanently paralysed and atrophied are usually cold; little blood circulates through them, for little tissue change takes place in them.

Diagnosis.—At the commencement of the attack—during the febrile stage of the disease—mistakes in diagnosis are easily made. The fever and constitutional disturbance are often thought to be due to teething, to the onset of one of the exanthemata (such as scarlet fever), of pneumonia, or cerebral meningitis. Where cerebral symptoms are prominent, as they sometimes are at the beginning of severe cases in young children, you can easily enough understand that a commencing attack of cerebral meningitis may be closely simulated. The vomiting, which in some cases of poliomyelitis anterior acuta is such a prominent symptom at the beginning of the attack, may lead the parents or even the doctor to think that the disturbance is merely due to stomach disorder. Further, the paralysis itself, when not very marked, may be overlooked and attributed, not only by the parents but occasionally too by the medical attendant, to simple muscular weakness and debility, the result of the fever and general disturbance.

But with the development and discovery of the paralysis—the *flaccid immobility* of the affected parts—the true nature of the attack becomes apparent.

Practically speaking, almost the only conditions which are

likely to present any real difficulty in diagnosis, once the paralysis is detected, are (1) an inflammation involving the motor cortical area of the brain; and (2) peripheral neuritis.

The differential diagnosis of poliomyelitis anterior acuta and of inflammation of the motor cortex of the brain. This condition, which is almost entirely confined to childhood, is rare—much less common than poliomyelitis anterior acuta. In cases of cortical inflammation, the resulting paralysis is usually (almost always) hemiplegic; the distribution of the paralysis in poliomyelitis anterior acuta is rarely hemiplegic; and in those cases of poliomyelitis anterior acuta in which one upper and one lower limb are paralysed, it is perhaps more common to find the lower limb on one side and the upper limb on the opposite side affected, than the upper and lower limbs on the same side involved; but this may occur. Again, although head symptoms (such as headache, vomiting, muscular twitchings, or even convulsions or actual coma) may occur in poliomyelitis anterior acuta, they are, comparatively speaking, rare, and when they do occur they are usually of brief duration and limited to the early stage of the illness; it is altogether exceptional for such symptoms to continue after the initial fever has subsided. Whereas, in the cerebral affection to which I am referring, head symptoms are prominent and they usually persist for some time after the febrile symptoms have completely disappeared. Frequently recurring convulsions, often unilateral and affecting the limb or limbs which subsequently become paralysed, are usually present. Another point of distinction is this, that cortical cerebritis or cortical phlebitis is apt to occur during, or immediately after, an attack of measles, whooping-cough, or other acute febrile disease. The same sequence is occasionally met with in cases of poliomyelitis anterior acuta, but it is very rare.

The difficulty in distinguishing poliomyelitis anterior acuta and cortical cerebritis occurs during the acute stage of the illness; any doubt which may remain in the mind of the practitioner as to the nature of the attack will be cleared up by the subsequent course of the case.

The condition of the affected (paralysed) limbs during the earlier periods of the case is quite different in the two diseases. In poliomyelitis anterior acuta, the lesion is situated in the lower division of the neuro-motor nerve apparatus; consequently,

rapid atrophy, the reaction of degeneration and abolition of the reflexes in the affected muscles are apt to result. Whereas, in cortical cerebritis, the lesion is situated at the very top of the neuro-motor apparatus; consequently, rapid atrophy, the reaction of degeneration and abolition of the reflexes do not result; in fact, in the course of time the reflexes in the paralysed limbs become exaggerated.

Both diseases may lead to the production of a permanently paralysed, atrophied and non-developed condition of a limb or limbs. But the muscles of the atrophied and non-developed limb, in cases of old-standing poliomyelitis anterior acuta, are flaccid, unless some of them, such as the calf muscles, should happen to be distorted by cirrhotic contraction. Whereas, the muscles of the atrophied and non-developed limbs, in cases of old-standing cortical cerebritis, are more or less rigid and contracted and their deep reflexes are exaggerated.

Patients affected with hemiplegia, the result of a cortical inflammation in childhood, are in after-life apt to suffer from epileptic convulsions which, in many cases, are unilateral and limited to the muscles of the paralysed side. Their mental development, too, is often interfered with. In some cases, a peculiar form of mobile spasm, to which the term athetosis has been applied, affects the paralysed limbs, especially the muscles of the forearm and hand. Whereas, an attack of poliomyelitis anterior acuta, although it frequently produces a permanent paralysis, which occasionally, though very rarely, is hemiplegic in distribution, does not interfere with the intellectual and cerebral development, and is not followed by any permanent cerebral symptoms (convulsions, athetosis etc.).

LECTURE VI

POLIOMYELITIS ANTERIOR ACUTA (*Continued*)

At the end of the hour yesterday, Gentlemen, I was speaking, you will remember, of the diagnosis and differential diagnosis of poliomyelitis anterior acuta. We saw that a difficulty in diagnosis is chiefly likely to arise at the commencement of the disease, before the characteristic paralytic symptoms have developed or have been noticed.

Once the paralysis is fully developed, there is rarely any difficulty in coming to a correct conclusion as to the nature of the case.

The recognition of poliomyelitis anterior acuta is based:—

Firstly, upon the presence of certain *positive* symptoms viz.:—The paralysis, the flaccid condition of the paralysed muscles, the rapid atrophy, and the presence of the reaction of degeneration.

These positive symptoms show that the lesion is situated in the lower division of the neuro-motor nerve apparatus, and, granting that it is spinal, in the anterior horn of grey matter.

And *secondly*, upon the presence of certain *negative* symptoms, viz., the absence of:—sensory disturbances (or at most the presence of very slight and temporary sensory disturbances), of paralysis of the bladder (though it must be remembered that there may be slight and temporary retention), of any affection of the rectum, and of bed-sores.

These negative indications show that the lesion is limited to the anterior horn.

We may say, then, that there are four steps in the diagnosis of a case of poliomyelitis anterior acuta, viz.:—

(1) The positive symptoms enable us to say (provided that peripheral neuritis can be excluded—I will refer to this point in detail presently) that there is a lesion of the anterior horn.

(2) The negative symptoms enable us to say that the lesion is confined (or for practical purposes confined) to the anterior horn.

(3) The rapid onset, which is usually attended with well marked fever, enables us to say that the lesion, which is confined to the anterior horn of grey matter, is an acute lesion.

(4) The facts that the acute lesion of the anterior horn of grey matter is attended with, and that its effects (the paralysis) are usually preceded by, fever, show that it is inflammatory in character.

The only lesions, other than poliomyelitis anterior acuta, which could possibly present such a combination of symptoms and conditions, are:—(a) Peripheral neuritis; (b) An ordinary acute myelitis and (c) A primary hæmorrhage which happened to be accidentally limited, as it were, to the region of the anterior horn.

But it is exceedingly rare to have these lesions (*b* and *c*) (ordinary acute myelitis and primary hæmorrhage) limited in this way. If they did happen to be so limited, they would be attended with exactly the same symptoms and signs as poliomyelitis anterior acuta. Fortunately, the distinction in the two last conditions—(*b*) and (*c*)—is of little practical importance, for the treatment in all three conditions (poliomyelitis anterior acuta, primary hæmorrhage and the ordinary form of myelitis limited to the anterior horn) is very much the same.

The differential diagnosis of poliomyelitis anterior acuta and of peripheral neuritis.—In some cases, it is extremely difficult, indeed it may be impossible, to distinguish poliomyelitis anterior acuta and peripheral neuritis. The distinction is especially difficult in those rare cases of peripheral neuritis, on the one hand, in which symptoms of sensory disturbance are entirely absent; and in those exceptional cases of poliomyelitis anterior acuta, on the other, in which muscular tenderness is a marked feature and in which there is some (slight) anæsthesia; further, it must be remembered that, in many of the latter cases, the inflammation of the spinal cord is probably complicated with an inflammation of the peripheral nerves.

In typical and well-developed cases, poliomyelitis anterior acuta can hardly be confounded with the ordinary typical form of peripheral neuritis; but all cases, as we have seen, are not

typical. I shall not occupy your time by going fully into the differential diagnosis on this occasion; for you will understand the points of distinction better after we have considered peripheral neuritis. But I may say, that the age at which poliomyelitis anterior acuta usually develops, the acute onset with fever, the rapid development of the paralytic symptoms, the absence of marked sensory disturbances, the fact that the paralysis is more localised and limited than it usually is in peripheral neuritis, and the absence of any definite exciting cause for a peripheral nerve inflammation (such as previous diphtheria, sore throat, alcoholic excess, etc.) together with the fact—and this is an important point—that there is no tenderness over the large peripheral nerve trunks, are the chief points on which we have to rely in order to distinguish the two conditions.

This difficulty in diagnosis seldom occurs in the child, for peripheral neuritis (if we exclude the diphtheritic form) is very rare in young children. I exclude mixed cases in which both poliomyelitis anterior acuta and peripheral neuritis are present. But even in older children and adults, provided that a clear history is forthcoming, the two affections are not likely to be confounded. The acute febrile onset and rapid development of the paralysis are, in the great majority of cases, alone sufficient to show the true nature of the case. It is the subacute forms of inflammation of the anterior horn of grey matter which are most likely to be confounded with peripheral neuritis. Further, in most cases of peripheral neuritis, sensory symptoms are present. In the rare and exceptional cases, such as that which I have already brought before your notice (see page 49), in which there are absolutely no sensory derangements, the diagnosis is most difficult; in some cases indeed impossible.

The differential diagnosis of poliomyelitis anterior acuta and of primary hæmorrhage, accidentally limited to the anterior horn of grey matter.—If a paralysis, clearly due to a lesion of the anterior horn, developed abruptly and instantaneously, without any previous constitutional and febrile symptoms, one would suspect primary hæmorrhage rather than inflammation; but, as I have already pointed out, before committing oneself to a diagnosis of primary hæmorrhage, an exceedingly rare condition, one must be absolutely satisfied that there were no premonitory symptoms

suggestive of previous inflammation. The slight febrile disturbance and constitutional symptoms, which attend a commencing inflammation of the anterior horn of grey matter, are easily overlooked; indeed, they frequently escape detection; and hæmorrhagic extravasation, usually slight, but in rare cases sufficiently extensive to give rise to symptoms, is common in poliomyelitis anterior acuta. In most cases, then, even when the sudden development of the paralysis suggests hæmorrhage, the extravasation of blood is more likely to be secondary than primary, i.e. the result of an inflammation which is insidiously developing.

The differential diagnosis of poliomyelitis anterior and of the ordinary form of myelitis accidentally limited to the anterior horn of grey matter.—In adults, even when the symptoms show that the lesion is chiefly located in the anterior horn, the presence of anæsthesia (and still more, of course, of paralysis of the bladder and of bed-sores) suggests that the inflammation of the anterior horn is due to the ordinary form of myelitis rather than to the special form of myelitis which we term poliomyelitis anterior acuta. As I have previously stated, there is reason to suppose that although both conditions are inflammations of the spinal cord, they are probably separate and distinct varieties. As I have already pointed out, it is probable that poliomyelitis anterior acuta is due to a definite and distinct irritant, possibly a micro-organism, and that it is, in respect to its etiology and nature, separate and distinct from every other form of inflammation of the spinal cord.

Although the diagnosis is usually easy, once the paralysis has developed, cases are occasionally met with in which a doubt may arise as to the exact nature of the attack.

The differential diagnosis of poliomyelitis anterior acuta and of reflex paralysis due to teething.—In children, paralysis occasionally occurs during teething, as the result, it is supposed, of reflex irritation. This reflex paralysis may easily be confounded with the much more serious paralysis due to inflammation of the anterior horn of the spinal cord. But true reflex paralysis of this kind is so rare that even if the child is teething it is well to hesitate before committing oneself to a positive opinion. If the paralysis entirely clears away in the course of a few days, it is allowable to conclude that it was probably reflex; for the

paralysis due to poliomyelitis anterior acuta very rarely indeed passes off with such rapidity; indeed, in cases of average severity, some permanent weakness usually remains; and in slight cases, the loss of power usually persists for some time. Fortunately (perhaps I should say unfortunately, for it is unfortunate for the patient) this difficulty in diagnosis rarely occurs. I recently saw a case in point. A healthy child, a year old, became feverish during teething, and in the course of a few hours manifested distinct loss of power in the right arm; the gums were lanced and a purgative and febrifuge were administered. In a few days, the fever subsided, the loss of power in the arm passed away and the child seemed well. After an interval of a week, a second attack of fever occurred and the right arm and leg became paralysed. The loss of power was now complete. The family physician, Dr. Willins of Penicuik, looked upon the case as one of reflex paralysis due to teething. Dr. Underhill, who was called in consultation, thought that it was probably due to poliomyelitis anterior acuta; and such was my own view when I subsequently saw the patient. But in the course of a few weeks the paralysis entirely disappeared, the leg recovering before the arm. I cannot positively say what was the exact nature of the lesion; it may have been a very slight attack of poliomyelitis anterior acuta; but if so, the hemiplegic disturbance and the very rapid and complete recovery were quite exceptional. Possibly Dr. Willins' opinion, viz., that it was a case of reflex paralysis due to teething, was correct.

The differential diagnosis of poliomyelitis anterior and of rickets.

—The muscular weakness and wasting due to rickets may at first sight and on superficial observation, be mistaken for the muscular weakness and wasting due to poliomyelitis anterior acuta. But the history (acute onset in poliomyelitis anterior acuta, slow and gradual in rickets), the distribution of the muscular weakness and wasting (localised in poliomyelitis, general in rickets), and the associated symptoms indicative of rickets (the presence of 'beading' of the ribs, the characteristic joint and bone alterations, the shape of the head, the sweating of the head, the muscular soreness, the sedate and sad expression, etc.) at once show the true nature of the case.

The differential diagnosis of poliomyelitis anterior acuta and diffuse muscular atrophy.—The history of an acute onset with fever

and with rapidly developed paralysis; the presence in the early stages of atrophy, and the reaction of degeneration; and the fact that, in the later stages, the atrophy is confined to groups of muscles and is not uniformly distributed over the whole or an extensive area of the muscular tissues of the body, are the chief points of distinction between poliomyelitis anterior acuta and a form of diffuse muscular atrophy which is occasionally, though very rarely, met with in young children.

A case of diffuse muscular atrophy in a child aged 18 months came under my observation at the hospital a week ago. I shall refer to it in detail when I come to speak of progressive muscular atrophy and pseudo-hypertrophic paralysis.

The diagnosis of poliomyelitis anterior acuta in old standing cases.

—The history and mode of onset of the attack are points of the greatest diagnostic importance in those cases in which the patient comes under observation at a late stage of the disease—i.e. when the paralysis and atrophy are of old standing.

But even in those cases in which it is impossible to obtain any definite information as to the early history and mode of onset, when, in other words, one has to form one's opinion from the present condition of the patient (from the observation of the results of the previous damage, so to speak), there is usually no difficulty in coming to a correct conclusion. In cases of old-standing poliomyelitis anterior acuta, the paralysis is not progressive. This fact alone enables one to distinguish the paralysis and atrophy due to poliomyelitis anterior acuta from the many other forms of muscular atrophy and paralysis with which, at first sight, it may appear to be similar.

The differential diagnosis of old-standing cases of poliomyelitis anterior acuta and of old-standing cases of cortical cerebritis has already been considered. (*See page 81.*)

Prognosis.—Let us now pass to the prognosis. Poliomyelitis anterior acuta is so very rarely fatal that it may be said to be unattended, or only in the most exceptional cases attended, with any danger to life. When the disease does prove fatal, it usually kills during the acute stage. Relapses are very rare, and recurrences (i.e. a return of the attack at some future period of life, after the first illness has been completely recovered from) are almost unknown. In the great majority of cases, once the

paralysis has reached its full development, some, and in most cases a considerable degree of, improvement may be expected to take place. Now, all of these are very satisfactory points. It is a great comfort to the parents to know, during the acute stage of the disease, that there is very little risk to life. During the acute stage of a disease attended with paralysis, the first thought of the parents is, Will the patient recover? Will the disease prove fatal? Laymen know in a general way that persons affected with paralysis often die. Laymen reason, 'This patient is suffering from paralysis, many people die from paralysis, therefore it is not improbable that our child who is suffering from paralysis will die.' But paralysis is a general, a wide term. We medical men know that in this particular form of paralysis the danger to life is very slight. We can therefore reassure the parents and can tell them that their child who is suffering from poliomyelitis anterior acuta will in all probability recover—will live. Such an opinion, when confidently expressed—and let me urgently beg of you, if you wish to succeed in practice, to endeavour to inspire confidence in your patients—is a great comfort to them.

But you can go further than this. When the parents' minds are relieved as to the immediate danger, they look to the future; the future progress of the case does not usually trouble them much until the first danger is past. When they are reassured as to the present, when they realise that the child is not likely to die, they begin to think of the future. They say to themselves, 'What a terrible thing it will be if our child remains permanently paralysed.' Fortunately, you can, in many cases, also give them comfort as regards the future. We have seen that, once the maximum intensity of the paralysis is reached, any alterations which occur are usually in the direction of improvement. If then, the paralysis is slight at the commencement, you can, with almost perfect certainty, assure them that it will to a very large extent pass off; if it is considerable, you can tell them that the worst is reached.

Another very important point. The public know that an attack of paralysis (rightly in the case of hemiplegia, due to vascular disease) is very likely to recur. They say, 'This is a case of paralysis; we know that after one attack of paralysis a second or a third, or it may be a fourth attack is apt to occur;

what a terrible thing it will be if our child has repeated attacks of paralysis.' But we have seen that relapses and recurrences in poliomyelitis anterior acuta are exceedingly rare. You can therefore reassure them again on this third point. You can confidently tell them that there is little or no risk of a relapse.

In trying to form an opinion as to the severity of the attack in any given individual case, the extent of the paralysis must of course be taken into account. When one limb only is affected, the prognosis is of course better than where two are involved. Remember that in most cases in which the initial paralysis is at all severe some permanent damage (paralysis) remains.

The most important questions which have to be determined for the purposes of prognosis are:—(1) What is the extent of the damage to the cord in the individual case under observation? (2) Is any considerable amount of paralysis likely to remain as a permanent condition? and (3) What effect will the disease be likely to have on the ultimate condition (future health and activity) of the patient?

During the first two or three weeks of the illness, a satisfactory answer to these questions can only be arrived at by observing the electrical condition of the affected muscle, and this, the information which the electrical test affords, is not perhaps always conclusive. During the first two or three weeks, the atrophy of the paralysed muscles is not sufficiently advanced and marked, even in severe cases, to guide the prognosis; it is only after six or eight weeks (when the wasting has become considerable) that one can with certainty say, from the size and bulk of the paralysed muscles, whether the paralysis is likely to be permanent or not. But at the end of the first ten days or a fortnight, the electrical examination, in many cases, gives most valuable information. The importance of the electrical test for prognosis can hardly be overrated. It is of course possible that some of the muscles which present the reaction of degeneration will completely recover, but the probability is all the other way, provided of course that the diagnosis is correct—that the case is really one of poliomyelitis anterior acuta. If the affected muscles fail to respond to the faradic current, we know that the attack is a severe one. Those muscles which present the reaction of degeneration (i.e. which fail to respond to the faradic current and which react in an abnormal—sluggish—way to the constant

current) will waste. Some of them or parts of them will, in all probability, remain permanently paralysed and atrophied. The extent, then, of the reaction of degeneration—or, to speak practically, the number of muscles which fail to respond to the faradic current and which present the sluggish (involuntary) form of contraction to the galvanic current—is, in the early stages of the disease, our most important guide to prognosis. You cannot, you will observe, form an accurate opinion as to the severity of the lesion from the mere fact that the limb is motionless, that its muscles are paralysed; for, as we have seen, the paralysis or some of the paralysis, which is present at the commencement of the attack, may be merely due to a temporary suspension of function, the result of œdema and inflammatory pressure or to destruction of axis-cylinders, nerve tubes and fine motor nerve fibrils. Provided that the nerve cells are not destroyed, there will not be permanent paralysis. The paralysis which is the result of shock, œdema and pressure, and of destruction of nerve tubes and axis-cylinder processes, will be recovered from in the course of time—always provided of course that the nerve cells (the trophic centres) connected with the affected nerve tubes and axis-cylinder processes, are not destroyed. But even in the early stages, the mere extent of the paralysis gives important information as to the severity of the attack. It is a rough guide, less certain than the electrical test, but still important. The more extensive and widely distributed the paralysis, the worse the prognosis; other things being equal, the more localised the paralysis, the better the prognosis.

The limb and the part of the limb which are affected must also of course be taken into account in forming an opinion as to the ultimate result in respect to deformity, activity, capability for special occupations, professions, etc. A paralysis of the right arm is, in right-handed people and older children, more important than a paralysis of the left. A severe paralysis of the arm, especially the left arm, is less serious than a severe paralysis of the leg. The deformity and impairment of activity which result from a wasted and paralysed arm are less than the deformity and impairment of activity which result from a wasted and paralysed leg. Paralysis and atrophy of the shoulder muscles is less serious than paralysis and atrophy of the muscles of the forearm; it is better to have a useless shoulder than a useless hand.

When one leg is affected, the deformity due to wasting and arrested development often becomes much more apparent about the time of puberty, in consequence of the rapid growth and development of the opposite (non-paralysed) limb which takes place at this time. I am often consulted in such cases, for the parents are naturally anxious to know whether something more cannot be done to promote the growth of the withered limb and to counteract the deformity of the leg and the twisting of the spine which are often considerable.

I have said that in the great majority of cases of poliomyelitis anterior acuta some improvement occurs, once the acute stage is passed. In individual cases, it is often difficult to predict the amount of improvement which will take place; but it is safe to say that so long as distinct improvement is going on, further improvement may still be hoped for.

When all the muscles of the affected limb or limbs present the reaction of degeneration, the prognosis is very bad. Under such circumstances, (provided always that the diagnosis is correct and that peripheral neuritis can be excluded) the patient will probably be more or less paralysed and crippled for life. If, at the end of from six or nine months, there is no return of motor power, no return of the faradic irritability, and no increase in the size of the paralysed and atrophied muscles, the paralysis and atrophy will probably be permanent. But you must remember that all the muscles of a limb rarely suffer in the same degree. Even if some (the more severely affected) muscles show no signs of recovery, considerable improvement in the condition of the limb as a whole may still take place in consequence of improvement in individual muscles and parts of muscles, which have been less seriously affected, and also in consequence of compensatory hypertrophy in those muscles of the limb which have escaped.

Again—and this is a most important point—because a muscle is severely affected—markedly paralysed and extensively atrophied—it does not follow that all of the paralysed and atrophied fibres will remain permanently affected. Some of the muscular fibres (those whose trophic nerve cells are not completely destroyed) may and probably will recover. The presence of atrophy and the reaction of degeneration does not necessarily show that the nerve cells are destroyed. These considerations show that time

and the progress of the case alone can show, with absolute certainty, what degree of improvement will take place.

Poliomyelitis anterior acuta does not interfere with the development of the sexual organs. This is a point about which (when the lower limbs are paralysed or partly paralysed) thoughtful parents are sometimes anxious. You can confidently reassure them on this point.

Two other questions which the parents are naturally anxious about are these:—Will the disease interfere with the intellectual development of the child? and Will the child when it grows up be more liable to suffer from nervous diseases, and especially from disease of the spinal cord, than other members of the community?

To the first question you can give an absolutely favourable answer. Poliomyelitis anterior acuta in no way interferes with the brain development.

With regard to the second point, we cannot perhaps speak quite so confidently. So far as I know, the subjects of old-standing poliomyelitis anterior acuta are not more liable to nerve diseases in after life (if we exclude chronic degenerations of the spinal cord) than other healthy persons. It is probable that degenerative processes (which often have their starting point in old lesions) are more likely to arise in a damaged than in a perfectly healthy cord, and a few cases have been recorded in which progressive muscular atrophy has developed in patients who in early life were affected with poliomyelitis anterior acuta; but this is not a matter of any great practical importance, for the probability of the occurrence of such degenerations is very slight. I know persons who are completely crippled—whose lower limbs are mere useless appendages, the result of an attack of poliomyelitis anterior acuta in early life—who are leading busy, intellectual and useful lives. Such persons are perhaps quite as likely to attain old age as other healthy members of the community; they are not exposed to some of the causes of disease which healthy people are (syphilis and injury, for example); they have therefore some, though it may not be great, compensatory advantages, to set against their grave disadvantages.

The question whether other children in the family are likely to be affected with the disease is sometimes put to one. On this point, you can reassure the parents, for the disease is rarely

hereditary and it rarely affects two members of the same family. It is well, however, to remember that exceptions to both of these statements are occasionally met with. On the mere theory of chances, it must occasionally happen that two children in the same family will be affected ; but the probability of such an occurrence is so small that it need not be taken into account. If two or more cases have occurred in any particular family, you will, of course, do well to give a guarded opinion as to the probability of other members of the family being affected. The occurrence of two cases in one family is suggestive that the members of the family have a special tendency to the disease, though it may of course be a mere accidental coincidence.

Treatment.—The facts that the paralysis is developed in the early stages of the attack and that it speedily attains its maximum degree of intensity are very unsatisfactory points, so far as treatment is concerned. In this disease, there is little hope of arresting the inflammation and of preventing the damage to the cord and the development of the paralysis. In the majority of cases, the damage is already done before we have an opportunity of applying treatment. Further, if one happens to see the case in the febrile stage, before the paralysis has developed, it is impossible to diagnose the disease, and therefore to employ effective treatment.

During the febrile stage of the disease, the indications for treatment are to reduce the fever and to allay the constitutional disturbance which is present.

Older patients should be confined to bed, but infants when they are ill, are usually better in their mother's or nurse's arms. The diet should consist entirely of milk and milk foods ; water of course may be given to allay thirst. A purge at the commencement of the attack is generally desirable ; an ordinary simple febrifuge mixture may be prescribed ; and such remedies as quinine, salicylate of soda, and antipyrin (if the fever is very high), in doses suited to the age of the child, may be given. Tartar emetic is a valuable remedy in many febrile affections, and especially in those cases in which the fever depends upon a local inflammation, but in this disease I prefer to give quinine at the beginning of the attack. When the fever is exceptionally high (104° or 105° F.) and especially when head symptoms are

present, cold-bathing and the application of cold to the head are advisable. If repeated convulsions should occur (but this is very rare), bromide of potassium, or bromide of potassium and chloral (in small doses) may be given.

With the development or discovery of the paralysis, the true nature of the attack becomes apparent, and the question at once occurs, Can anything be done to arrest the inflammation and to prevent the destruction of the anterior horn of grey matter? Although, in most cases, the damage is already done when the physician first sees the patient, local measures should in all cases be employed for arresting the inflammation, when the case is seen sufficiently early. Leeches or cups applied over the affected part of the spine (cervical or lumbar enlargement of the cord) are probably the most effective means of treatment. A fly blister in some cases seems to be useful. Warm fomentations are recommended by some physicians; cold applications in the form of an ice-bag to the spine, by others. I need not discuss the relative advantages of heat and cold (applied locally) for the arrest of the vascular changes which occur at the beginning of an inflammation. The question is not new to you. We have considered it more than once in the previous part of the course. But I may say that it is not an easy matter to apply an ice-bag to the spine of a young child, even if you think it desirable to do so. There is a practical difficulty in carrying out the treatment which it is well to realise. A young practitioner who is called to a case of poliomyelitis anterior acuta and who has been taught to believe in the local application of cold, but who has never seen an ice-bag applied to the spine, gives instructions that the child should be made to lie on its face and an ice-bag continuously applied. The mother or nurse shakes her head; she knows that it will be no easy matter to carry this prescription into effect; and the young doctor falls considerably in her estimation in consequence; she sees that he is not a practical man. Of course older children and adults can be made to lie on the face for hours together; but the position is neither a pleasant nor a comfortable one.

With the object of preventing the blood gravitating to the affected part of the spinal cord it is in most cases probably sufficient to make the patient lie on his side. The ice-bag can be satisfactorily applied in this position; the ice should be frequently

changed, care of course being taken that as it melts the water does not escape and wet the patient.

By attention to position (keeping the patient off his back) and the continuous application of local cold to the spine, the vascular and inflammatory changes in the cord can no doubt be, in some degree, restrained. It is, I think, probable that the local application of hot fomentations is quite as effective as the ice-bag, and in most cases the warm applications are more agreeable to the patient. It is important to remember that the sensations of the patient are, in many diseased conditions, a valuable guide to treatment. As a general rule, patients like what is good and dislike what is bad for them. Of course this rule does not always apply. In disease, as in health, the things which we like and desire are not always the most suitable and the most beneficial. But speaking generally, those local applications which are most grateful to the patient are the most beneficial. In the case of children, and the vast majority of cases of poliomyelitis anterior acuta occur in young children, I prefer warm fomentations to the ice-bag. But dry cupping or leeching is probably the most effective means of local treatment at this, the early, stage of the disease.

Ergot and belladonna have been strongly recommended by some authorities in the early stages of poliomyelitis anterior acuta, with the object of producing contraction of the blood vessels of the inflamed part; but it is doubtful whether they exert any material influence upon the course of the disease.

We may say, then, that the treatment of poliomyelitis anterior acuta in the early stages is practically the same as that of any other form of local inflammation.

After the fever has subsided, the main objects of treatment are to promote absorption of the inflammatory exudation and to favour the recovery of the damaged nerve elements. The affected parts of the cord should, so far as possible, be kept at rest; everything which is likely to irritate and throw a strain upon the damaged nerve elements should be avoided; strychnine, electricity, and vigorous attempts to produce voluntary movements are, at this stage, apt to do harm rather than good.

The general health should, so far as possible, be kept in the highest possible state of efficiency, and the condition of the bowels should be carefully regulated. The sick-room should be

well-ventilated ; during the febrile stage, plenty of fresh air is desirable ; care should be taken that the room is not too hot ; even in winter, it is a great mistake to have the bed-room overheated.

During this, as during the febrile stage of the disease, quinine may, I think, be given with advantage in small doses ; the syrup of the iodide of iron and iodide of potassium may be given with the object of promoting absorption of the inflammatory products. In some cases of poliomyelitis anterior acuta, iodide of potassium seems to produce depression. In such cases, the syrup of the iodide of iron or hydriodic acid are preferable. Mercury, given by the mouth or better still by inunction, has also been recommended at this stage of the disease, but it is of doubtful efficacy.

After a month, the more acute changes may reasonably be expected to be at an end. Provided there is no indication to the contrary, the child may now be taken out-doors.

After the first month or six weeks, cod-liver oil (if the stomach will digest it), the syrup of the phosphates, arsenic and strychnine are probably the most important drug remedies which we possess.

During the first month, the affected limb may be rolled up in cotton wool, but it should not be heavily weighted ; the object is to keep the paralysed parts at an equable temperature.

Until the acute changes have subsided, strychnine and electricity should not be employed. This is an important statement. Many practitioners when they meet with a case of paralysis at once think of strychnine and electricity. The presence of paralysis seems always to suggest in some men's minds the necessity of prescribing electricity and strychnine, just as a cardiac murmur seems always to suggest in some men's minds the necessity of prescribing digitalis. Such a routine method of prescribing is no doubt in some cases attended with benefit, but in others it is harmful. I am of opinion that electricity and strychnine should be avoided in the early stage of poliomyelitis anterior acuta.

At this stage, gentle massage of the affected limb is the only local treatment which is desirable, and care should be taken that the massage is not too forcibly and vigorously applied. Massage, like every other remedy, requires to be employed with

judgment and caution. Professional rubbers often rub far too vigorously. It is necessary to remember this. Now-a-days all sorts of persons are adopting 'rubbing' as a profession. Some of them have brains, some have not. Some women seem to take to rubbing simply because they are big muscular women. If you put one of these big muscular women without brains, to rub a young child who is just recovering from an attack of poliomyelitis anterior acuta, the rubbing will be likely to do harm rather than good.

LECTURE VII

POLIOMYELITIS ANTERIOR ACUTA (*Continued*)

At the end of the hour yesterday, Gentlemen, I was speaking, you will remember, of the treatment of poliomyelitis anterior acuta. We saw that although, in most cases, it is impossible to arrest the inflammation in its early stage and to prevent the development of paralysis (owing to the fact that the destruction of the nerve elements has already taken place when the paralysis is detected and the nature of the case is apparent) an effort should be made (provided the case is seen sufficiently early) to restrain the inflammatory change and limit the damage to the nerve cells. I described to you the chief therapeutic means which it is advisable to employ in the endeavour to carry this indication into effect. I then told you that after the acute stage of the disease is passed the affected part of the cord should, so far as possible, be kept at rest; that every source of irritation should be avoided; that the health of the child should be maintained in the best possible state of efficiency; and that remedies should be administered with the object of promoting absorption of the inflammatory products.

Further, we saw that when four to six weeks have elapsed, when, in other words, it is reasonable to suppose that the acute changes have subsided, the great object of treatment is to aid the restoration of the damaged nerve elements.

With this object, the child should be encouraged to move the affected (paralysed) parts. In all cases of paralysis the best stimulus which can be employed is the normal stimulus of the 'Will'; it excites the functional activity of the nerve cells and aids the restoration of the damaged (degenerated) nerve fibres; each fresh effort of the 'Will' sends a nerve impulse up to the point of the lesion; repeated efforts of the 'Will' gradually, as it were, force a passage through the 'block.' Frequently repeated, voluntary stimuli are especially useful in those cases in which

the paralysis is due to a lesion of the nerve tubes; but I am satisfied that in every form of paralysis they are the best stimuli which can be employed.

After the acute changes have subsided, massage and electricity are most useful and important means of treatment. At the end of the last lecture I took occasion to emphasise the fact that the massage must be judiciously and carefully employed. The same statement applies to the use of electricity. Most of the patients who are affected with poliomyelitis anterior acuta are young children, and it is not always an easy matter to apply electricity to a young child.

Our object in using electricity in this disease is twofold, viz., (1) to maintain the nutritive condition of the affected muscles; and (2) to restore (or aid the restoration of) the damaged nerve elements and degenerated muscles.

Some authorities recommend that the electric current should be passed through the cord with the object of promoting absorption of the inflammatory products; but, as I have already told you, I am of opinion that, during the early stages of the disease, electricity in any form (whether applied to the cord itself or to the paralysed muscles) is inadvisable; during the acute stage electrical treatment is apt, I think, to do more harm than good.

Other authorities recommend that after the acute changes have subsided, the constant current should be passed through the cord, one electrode placed on the back at the seat of the lesion, the other over the front of the abdomen or some other part of the spinal column. It is claimed that electricity employed in this way stimulates the spinal cord and promotes recovery of the damaged nerve elements (cells and fibres). There can be no doubt that the so-called catalytic action of the constant current is of importance. But the information which we possess as to the therapeutic effects of the constant current, when used in the manner which I have just described in cases of poliomyelitis anterior acuta, is very indefinite. Further, you must remember that if the current is too strong, it may do harm rather than good; and it is very doubtful if a very weak current applied over the spine reaches the spinal cord. Some good authorities are so doubtful of the beneficial results which electricity produces when applied in this way, and are so convinced of the prejudicial effects which a too powerful current may

produce in cases of poliomyelitis anterior acuta, that they discountenance the application of electricity to the spinal column. For my own part, I am disposed to think that a weak current cannot do any harm, but I am not prepared to say that it will do good.

The direct application of electricity to the affected (paralysed) muscles and nerves is much more important. One pole of the battery may be placed (fixed) over the motor point—the point of entrance of the motor nerve—the other may be moved from point to point over the body of the muscle. The position of the motor points may be ascertained by referring to some one or other of the published diagrams or charts. Students cannot be expected to remember the position of these motor points. For my own part I do not profess to remember them. If I wish to apply electricity to a motor point the exact position of which I do not remember, I refer to the chart. When you enter practice and have to treat cases of paralysis, you will be able to adopt the same plan. You are not allowed to refer to a chart of the motor points in examinations; and I am strongly of opinion that you should not be expected to remember, and that you should not be examined on, the position of these motor points. I do not advise you, either as students or practitioners, to attempt to burden your memories with these details—useless details of this kind as I think them. You will find that it is difficult enough to remember the essential and important things. They are—at least I find them so—quite enough. There is a great deal too much made of minute details of this kind in lectures and examinations. Some examiners seem to think that it is their business to try and puzzle the student. The business of an examiner is to find out what the student knows, not to try and find out what he does not know. The object of examinations (putting honour examinations of course out of account) is to see if the candidates are fit to enter practice. Not one practitioner out of a thousand knows or attempts to remember the position of the motor points. The student, therefore, should not be expected to know and remember them. The examiners who examine on these minute and, as I think, quite unnecessary details probably in some instances at all events have to cram them up the night before. Such men are not fit to be examiners.

But to return to the application of electricity to the paralysed

muscles and nerves. The object of electrical treatment is to maintain the nutrition of the muscles which are less severely affected and to help to restore the functional activity of the muscles and nerves which are severely damaged (degenerated).

The question naturally arises, Which form of current (the interrupted or the constant) should be employed. Both are useful. The muscular fibres which are undergoing the rapid form of atrophy and which present the reaction of degeneration will not respond to the interrupted current; if, then, you wish to excite their functional activity, the constant current must be used. The muscular fibres which are less severely damaged, and those fibres of the paralysed muscles which are unaffected or only temporarily affected, will respond to the interrupted current; to excite their functional activity, the interrupted current is all that is required.

Further, you should remember that a slowly-interrupted constant current is less painful than the quickly interrupted faradic current. In the case of children, this is a most important consideration. Children are easily frightened and agitated, and if you begin by applying a strong current or a faradic current to a child, you will give it pain, frighten it, and make it cry. At the next visit, it will probably begin to cry immediately it sees you or the electrodes; the agitation which is produced at each visit will certainly not be beneficial to its weakened nerves and muscles; indeed, electrical treatment under such circumstances will be likely to do much more harm than good. And here let me say that the object of the physician should be to produce a pleasant and stimulating, not a terrifying and depressing, effect, upon the minds of his patients. A good bedside manner, and (what is far more important and valuable) a real, honest, kindly and sympathetic interest in the patient and his sufferings, not only enables the practitioner to succeed in making and keeping a practice, but it actually helps his patients to recovery. There is much more in mental therapeutics than some people would seem to allow or to suppose.

It is essential, therefore, to endeavour to apply the electricity in such a way that it is neither disagreeable nor painful to the child. It is always advisable to begin with a very weak, slowly-interrupted current. In the case of young and nervous children, it is a good plan to instruct the nurse to sponge the paralysed

limbs with the electrodes only (i.e. without any electricity) several times a day in order that the child may get accustomed to them. After a few days, you can begin to use a slowly interrupted and very weak current. At each succeeding visit, the strength of the current may be slightly increased until the maximum strength of current, which can be pleasantly and satisfactorily borne, is reached. There you should stop. If the electricity is to be beneficial, it must of course be sufficiently strong to excite the functional activity of the paralysed muscles; a current which is too weak to produce muscular contractions is not likely to do much good.

The stable (fixed) electrode should be of large size.

In the case of older children and adults, the faradic current may be used, provided that the muscles respond to it.

Systematic electrical treatment and massage should be diligently continued not only for weeks, but for many months—until, in fact, the practitioner is satisfied that the improvement is at an end.

During the future progress of the case, the general health should be carefully attended to. If the weather is warm, the child may be allowed to spend a large part of its time in the open air, care being taken that it neither gets overheated nor overchilled. During this stage, nerve tonics should be administered. Strychnine, which is contra-indicated during the earlier periods of the case, is, I believe, a useful remedy at this period of the case.

In the case of children—and most of our patients who are suffering from poliomyelitis anterior acuta are young children—the drug is, I think, best given by the mouth. Half a drop of the liquor strychnine or one drop of tincture of nux vomica, three times daily, is a sufficient dose for a child of a year old, at all events to begin with. Some authorities advise that the remedy should be given hypodermically; but young children strongly object to the prick of the hypodermic needle; the daily repetition of the prick disturbs and upsets them and makes them dislike the doctor. I am not satisfied that these disadvantages are counterbalanced by the advantage which the hypodermic method of administration gives. If the hypodermic method is employed, half a drop of the liquor strychnine may be given *once daily* to a child of a year old, by the hypodermic method.

In all cases, the effects of the remedy, whether it is given by the mouth or hypodermically, should be very carefully watched.

Arsenic is another valuable nerve tonic; it is generally well borne by children. If the child is anæmic, iron may be given in addition to the arsenic. Cod-liver oil and quinine are also beneficial. Cod-liver oil is chiefly useful in delicate, badly nourished children, and during cold weather.

At the end of a year or eighteen months, active treatment may usually be suspended. The muscles which are going to recover will probably have recovered before this time. Any further improvement must be the result of natural growth and development. Massage may still be continued. If the parents can afford it, the services of a skilled and trained *masseuse* may be employed; but the nurse or mother can, after a few lessons and with a little practice, usually do all the rubbing which is required. It will not, of course, do the child any harm to continue the electricity, or to carry out a course of electricity once or twice a year, if the parents desire it and can afford it.

In most cases, the chief improvement is likely to take place, and is chiefly attained, during the first nine or twelve months. After this period, the main indications for treatment are to attend to the general health, to aid the muscular development, and to prevent deformity.

It is very important to prevent the joints of the paralysed limb becoming fixed in a vicious position or distorted in the manner which has been already described (see page 78). With this object, mechanical appliances, such as elastic bands fixed to the affected limb in such a way that they pull in the direction of the paralysed muscles, may be applied, and passive movements of the affected joints should be systematically performed.

I may say in passing that the prevention of contractures and deformities is a very important point in the treatment of all forms of paralysis and especially in those cases in which the paralysis is the result of peripheral nerve lesions. I shall refer to this point more in detail when I come to speak of alcoholic paralysis.

With the object of keeping a weak joint or limb fixed in a proper position and of overcoming and preventing contractures and deformities, mechanical supports have in some cases to be employed. As the child gets older, the need for these mechanical appliances often becomes greater. At or about the time of

puberty, the disparity between the two limbs, when one only is affected, usually becomes much more marked. At this age, the patient is often brought to the physician to see if something more cannot be done. Fortunately, in many cases a good deal can be done by means of suitable mechanical appliances to correct or prevent deformity, to lessen the limp, to aid the walking power, etc. The chief use of mechanical appliances in such cases is, either to support a part which is weak (the ankle joint for example, which is often so weak that it turns in when the child stands upon it), or to give height (length) to a shortened limb. It is very essential to see that any mechanical support which you recommend is not too heavy. It is usually the case that some of the muscles which have been severely damaged, although very weak, still retain some functional activity; they may be still able to move the withered limb. The muscles of the thigh, for example, often retain some power, when the muscles below the knee are completely or almost completely paralysed and atrophied. It is very essential to take care that you do not overload these weakened muscles. If you overweight the limb, you handicap the enfeebled muscles so heavily that you do more harm than good; you hamper their activity, prevent their growth and development and increase the difficulty in walking rather than lessen it. In choosing a mechanical support, the object should be to give as much support to the weakened joints as possible, and to overweight the weakened muscles as little as possible. Although by means of suitable mechanical supports the walking power can often be improved and deformity lessened or prevented, there is of course at this stage of the disease, no hope of removing the paralysis; and yet this is just what some people seem to profess to be able to effect. Since it is impossible to restore the atrophied and paralysed muscles or to promote in any material degree the growth of shortened and undeveloped bones, it is dishonest in old-standing cases of poliomyelitis anterior acuta to encourage the parents to hope for great things. I do not mean to say that treatment is altogether ineffectual at this stage of the case. In many cases, systematic massage, electricity, and gymnastics undoubtedly produce some improvement; and even a little improvement may be most useful and valuable. But it is downright quackery to hold out hopes of a substantial degree of improvement in such cases, from any plan of treatment, however

assiduously and continuously employed. Unfortunately, those of us who deal honestly with our patients in this respect often lose them. They come to us hoping that something effectual can be done. If we are honest, we tell them that at this stage of the disease it is impossible to remove the paralysis, or to effect any substantial improvement, but that some improvement may possibly be effected by suitable treatment (massage, electricity, mechanical appliances, etc.) In many cases they are dissatisfied with this opinion and they go to some one else—perhaps to a properly qualified medical man, perhaps to a professional rubber—who perhaps encourages them to hope for a much greater degree of improvement than can possibly take place. After several weeks or months of treatment, the patient is sent home very much *in statu quo*; and told to come back for further treatment in the course of a short time. This process of fee-extraction is repeated again and again—as long in fact as the parents will submit to it. After a year or two, and sometimes before this, they usually have had enough of it; they see that the opinion of the first man was correct; but the second practitioner has in the meantime reaped his reward; he has got as much as he could out of them, and that was what he meant to do. Let me beg of you, gentlemen, to do your best to discountenance and suppress such discreditable modes of practice.

In some cases of old-standing poliomyelitis anterior acuta, surgical procedure is helpful. Division of the tendo Achillis is advisable in some cases in which the muscles of the calf are cirrhosed and the heel drawn up by the contracted and atrophied calf muscles. But here let me say that the tendo Achillis is often cut quite unnecessarily. In many cases in which the growth of the leg is arrested, the patient walks on the toes with the heel drawn up from the ground. On superficial observation, the drawing up of the heel may appear to be due to contraction of the calf muscles, whereas it is merely the result of shortening of the leg. Section of the tendo Achillis is not required in these cases. The proper way to correct the deformity is to thicken the sole of the boot and to increase the size of the boot-heel.

Resection of the knee joint has been recommended in some cases of poliomyelitis anterior acuta, in which (in consequence of the paralysis and atrophy and loss of muscular support) the knee joint hangs loose like a flail, but in which the muscles

which act on the hip joint are capable of acting on the limb as a whole.

So much, then, for the first system disease of the spinal cord. The consideration of poliomyelitis anterior acuta has occupied a great deal of time, but the time will not be misspent; for many of the points which I have brought before you in the preceding lectures have an important bearing upon other diseases of the nervous system which we shall have to consider in the subsequent parts of the course.

SUBACUTE AND CHRONIC INFLAMMATION OF THE ANTERIOR HORN OF GREY MATTER

POLIOMYELITIS ANTERIOR SUBACUTA AND CHRONICA¹

Introductory Remarks.—Cases are occasionally met with in which paralysis and muscular atrophy are developed more gradually than in poliomyelitis anterior acuta, and in which there seems reason to suppose that the lesion is situated in the anterior cornual region of the spinal cord. There is, however, still much uncertainty with regard to the exact pathology of some of these cases. In many of the cases, which were formerly supposed to be due to subacute or chronic inflammation of the anterior horn of grey matter, the lesion is undoubtedly situated in the peripheral nerves rather than in the spinal cord. I have already told you that in rare cases of peripheral neuritis the motor fibres are chiefly, indeed it may be alone, involved. Now in the rare cases of peripheral neuritis in which there are few, indeed it may be no, sensory symptoms, the diagnosis is attended with great difficulty; and the resulting paralysis and muscular atrophy are highly suggestive of a lesion of the anterior horn of grey matter. I have previously referred to a case in point (see page 49). In another case which came under my notice some three years ago, and which at first sight appeared to be a case in which the lesion was spinal and situated in the anterior horn of grey matter, the diffuse and symmetrical character of the atrophy and the presence of slight tenderness over the nerve trunks were the only means of arriving at a correct diagnosis. They suggested that the lesion was situated in the peripheral nerves rather than in the spinal cord. The correctness of this opinion was proved by the subsequent course of the case, for after many months the muscular atrophy, which was at first almost complete, was in large measure recovered from. I shall refer to

¹ Before studying this subject, the reader is advised to read the lectures on progressive muscular atrophy.

this case in more detail when I come to speak of peripheral neuritis.

It is not improbable, I think, that in some cases of the so-called '*peroneal type of muscular atrophy*,' the lesion is a chronic inflammation of certain segments of the anterior cornual region of the spinal cord, rather than an inflammation of the peripheral nerves. But this again is a disputed point.

One must be on one's guard, then, against mistaking cases of peripheral neuritis for cases of subacute inflammation of the anterior horn of grey matter. But although it may be granted that in many cases in which subacute or chronic inflammation of the anterior horn of grey matter is supposed to be present, the lesion is in reality situated in the peripheral nerves, it must, I think, be admitted that undoubted cases of poliomyelitis anterior subacuta and chronica are occasionally met with.

POLIOMYELITIS ANTERIOR CHRONICA

(CHRONIC INFLAMMATION OF THE ANTERIOR HORN OF GREY
MATTER)

Poliomyelitis anterior chronica is characterised by the slow and gradual development of paralysis and muscular atrophy. It is not improbable that in some rare cases of diffuse muscular atrophy which are usually considered to be myopathic in origin, the lesion is spinal (a chronic inflammation of the anterior cornual region). Further, some authorities suppose that a chronic inflammation of the anterior horn of grey matter is the pathological substratum of the ordinary (Aran-Duchenne) form of progressive muscular atrophy; others, however, think that in that disease the destruction of the multipolar nerve cells of the anterior horn is due to a degenerative rather than an inflammatory process.

Degeneration and chronic inflammation of the anterior cornual region will necessarily produce very similar symptoms, viz., a slowly developed muscular atrophy and paralysis. But, so far as we know, a chronic inflammation of the anterior cornual region does not affect the segments of the cord in the same serial order, so to speak, in which they are involved in typical cases of progressive muscular atrophy. In the great majority of cases of

the ordinary spinal form of progressive muscular atrophy, some of the segments of the cervical region are first affected, and the atrophy and muscular weakness are usually first developed in the small muscles of the hand. In poliomyelitis anterior chronica, the grey matter of the lumbar enlargement seems in most cases to be first affected; the resulting atrophy and muscular paralysis are consequently first observed in the lower extremities. Further, in poliomyelitis anterior chronica, the atrophy is usually more diffused than in the ordinary typical form of progressive muscular atrophy. In that disease, individual nerve cells are picked out and slowly destroyed, and the individual muscular fibres with which they are connected undergo a slow and gradual atrophy. Whereas in poliomyelitis anterior chronica, groups of nerve cells are simultaneously affected and larger masses of muscle simultaneously become paralysed and undergo the atrophic process.

In some cases which appear to belong to this group (poliomyelitis anterior chronica), almost all the voluntary muscles of the body are affected simultaneously or in rapid succession. This seemed to be the course of the disease in a case to which I will refer in more detail when I come to speak of the myopathic forms of progressive muscular atrophy.

Cases are still more frequently met with in which localised lesions of the anterior cornua are developed in the course of other chronic diseases of the spinal cord. This is notably the case in amyotrophic lateral sclerosis (which, as we shall afterwards see, is by some observers regarded as identical with the ordinary Aran-Duchenne type of progressive muscular atrophy) and in syringomyelia. Localised muscular atrophy due to a lesion of the anterior cornu is occasionally met with in locomotor ataxia, spastic paraplegia, leprosy, diabetes mellitus, joint disease, etc. In the latter case, the muscular wasting is apparently the result of a reflected lesion (irritation) originating in the diseased joint and conducted through the sensory nerves and posterior roots to the multipolar nerve cells of the anterior horn. In some of these cases—probably in most cases of locomotor ataxia and diabetes mellitus—the muscular atrophy and resulting paralysis are due to an inflammation of the peripheral nerves rather than of the spinal cord; but in other cases there seems to be no doubt that the lesion is situated in the anterior horn of grey matter.

SUBACUTE INFLAMMATION OF THE ANTERIOR
CORNUAL REGION

(PARALYSIE GÉNÉRALE SPINALE ANTÉRIEURE SUBAIGUË)

Cases are occasionally met with in which the lesion seems to be a subacute inflammation of the anterior horn of grey matter. The leading symptom which characterises this group is paralysis followed by (not preceded by) muscular wasting. The paralysis is developed more gradually than in cases of poliomyelitis anterior acuta; and the onset may be, and often is, entirely unattended with febrile and constitutional symptoms. These cases seem to be identical with the condition which Duchenne described under the term, *Paralysie générale spinale antérieure subaiguë*. This is a rare affection which, so far, has been almost exclusively observed in adults between the ages of thirty and fifty; but cases are occasionally met with in children. In some of its features, it closely resembles the so-called 'peroneal type of muscular atrophy.'

Pathology.—In most of the cases which have as yet been examined post-mortem, appearances indicative of chronic inflammation in the region of the anterior cornua were present; the multipolar nerve cells were more or less extensively destroyed; the vessels dilated; the connective tissue thickened; and the nuclei and Deiters' cells more prominent and more abundant than in health.

In other cases, which were considered to belong to this group (but which were perhaps cases of peripheral neuritis), no marked lesion in the anterior cornua of the spinal cord seemed to be present. In order to explain these cases, Erb has suggested that degeneration of the peripheral nerves and muscles may, in some cases, perhaps result from a functional derangement of the multipolar nerve cells in the anterior cornua (the trophic centres for the muscles), or from structural changes in the nerve cells which are too minute to be recognised by our present means of observation.

Etiology.—Nothing definite is known as to the causes of the condition. Erb has suggested that lead poisoning is perhaps the

cause in some cases. In support of this view it must be allowed that paralytic symptoms, closely resembling those which occur in subacute inflammation of the anterior horn of grey matter, do sometimes result from plumbism; and further that in some cases of lead poisoning inflammatory changes have actually been found in the grey matter of the anterior horn.

Clinical History.—As a rule, the onset is more or less insidious and gradual. In some cases, premonitory symptoms, consisting of aching pains in the back and limbs, a feeling of excessive weariness and muscular weakness, or slight febrile disturbance, are observed.

The characteristic symptom is motor weakness, which gradually increases until complete paralysis is reached. In the majority of cases, the paralysis begins in the lower extremities; the muscles of the leg are usually first invaded; then those of the thigh; and finally those of the hip. This is the *ascending type* of Duchenne.

After the lower extremities have become paralysed, the upper limbs are in turn involved, the extensor muscles of the fingers being in many cases the first to suffer. The muscles of the upper arm and shoulder are subsequently affected. The muscles of the trunk may also become involved.

In exceptional cases the paralysis first affects the muscles of the upper extremities, and then extends to the lower limbs. This is the *descending type* of Duchenne.

In rare cases, the muscles on one side of the body are much more severely affected than those on the other.

The paralysis usually presents all the characteristic features due to a lesion of the anterior horn of grey matter. The muscles become markedly atrophied and in many cases present the reaction of degeneration in a more or less typical degree; the reflexes are at first diminished, and soon completely abolished.

It is important to note that the paralysis is the first event; in other words, the atrophy follows the paralysis. This fact serves to distinguish the condition from progressive muscular atrophy, in which the muscular wasting precedes, or is produced simultaneously with, the muscular weakness.

The sensory functions are practically intact. Slight numbness may be present, but there is never marked anæsthesia.

The functions of the bladder and rectum are not interfered with.

Course.—The rapidity with which the paralysis extends and the disease advances varies in different cases. The muscles first affected become quickly paralysed and then wasted; but the maximum extent (diffusion) of the paralysis may not be reached for months, or even, it is said, for years.

After the paralysis has reached a certain degree of development, a stationary period generally occurs; but in some cases, the lesion continues to extend, and ultimately reaches the upper part of the spinal cord and the medulla oblongata. Bulbar symptoms are then developed, and death results from respiratory complications.

In many cases, improvement occurs after the stationary period has lasted for a few weeks or months. Recovery usually takes place in the reverse order to the mode of onset; in other words, the muscles which are last involved are the first to recover. In exceptional cases, the recovery is complete; but in most cases, some paralysis remains.

The total duration of the disease is usually, it is said, from one to four years.

Diagnosis.—The symptoms seem to show that the lesion is situated in the anterior cornual region of the spinal cord rather than in the peripheral nerves; but, as I have already pointed out, the differentiation from peripheral neuritis is in some cases very difficult. The facts that the paralysis is the first event; that the muscular atrophy follows the paralysis; that there is practically an entire absence of sensory symptoms; together with the peculiar way in which the paralysis and atrophy extend (one group of muscles or muscular mass being first paralysed and then atrophied, and other groups or masses of muscle being subsequently affected *seriatim* in the same manner—first paralysed and then atrophied) are the most important points from a diagnostic point of view.

Poliomyelitis anterior subacuta has to be distinguished from poliomyelitis anterior acuta, progressive muscular atrophy and lead palsy.

The differential diagnosis of subacute inflammation of the anterior cornu and poliomyelitis anterior acuta.—Subacute inflammation of

the anterior horn of grey matter usually occurs in adults between the ages of thirty and fifty, rarely in children; while, poliomyelitis anterior acuta very rarely indeed occurs in adults and is almost entirely confined to childhood. In subacute inflammation of the anterior horn of grey matter, the onset is, as a rule, gradual and unattended by fever and cerebral symptoms; while, in poliomyelitis anterior acuta, the onset is abrupt, fever and constitutional symptoms are usually prominent, and cerebral symptoms are not uncommon. In subacute inflammation of the anterior horn of grey matter, the paralysis extends from point to point, usually from below upwards; the maximum extent (as regards distribution) of the paralysis is never at once reached; while, in poliomyelitis anterior acuta, the maximum degree and distribution of the paralysis are reached quite at the early stage of the attack, and any changes which subsequently occur are in the direction of improvement. In subacute inflammation of the anterior horn of grey matter, the result is, in some cases, recovery; in others, death. Even in those cases in which the paralysis and atrophy are extensive and marked, complete recovery occasionally occurs. When the disease proves fatal, death usually occurs after two, three, or four years, and is in many cases due to extension of the lesion to the medulla oblongata and the production of bulbar symptoms. While, in poliomyelitis anterior acuta, some paralysis usually remains if the case is at all severe; and even when the attack is by no means very severe, complete recovery is rare. Poliomyelitis anterior acuta is very rarely fatal; and when death does occur, it almost always takes place during the acute stage of the disease.

In short, the mode of development of the paralysis and the course which the disease pursues are entirely different in the two cases.

The differential diagnosis of subacute inflammation of the anterior horn of grey matter and of progressive muscular atrophy.—In subacute inflammation of the anterior horn of grey matter, the paralysis usually begins in the lower extremities; while, in the great majority of cases of (typical, spinal) progressive muscular atrophy, the muscles of one upper extremity (usually the small muscles of the hand) are first affected. In subacute inflammation of the anterior horn of grey matter, whole muscles or groups of muscles are attacked simultaneously; the muscles are first

paralysed and then atrophied; the reaction of degeneration is usually present in a more or less typical form, and the reflexes belonging to the affected muscles are abolished at an early stage of the case. While, in progressive muscular atrophy, whole muscles are never affected all at once, but individual fibres and groups of fibres are slowly and gradually destroyed; further, the wasting is the first event, or at all events the loss of muscular power is developed simultaneously with, and in direct proportion to, the muscular atrophy. The reaction of degeneration is rarely if ever present in a typical and fully-developed form, and the reflexes are not abolished until a considerable amount of muscular wasting and paralysis have been established. In subacute inflammation of the anterior horn of grey matter, the paralysis in many cases undergoes improvement, and recovery is in some cases complete; while in progressive muscular atrophy the atrophy and paralysis tend markedly to advance; muscles which have once become atrophied are rarely if ever completely restored.

The differential diagnosis of subacute inflammation of the anterior horn of grey matter and of lead poisoning.—There is of course no difficulty in distinguishing the typical form of lead paralysis (wrist-drop) from the disease which we are now considering. Chronic lead poisoning does, however, sometimes produce a more generalised form of muscular wasting which very closely resembles, and in some cases is perhaps identical with, the paralysis due to a subacute inflammation of the anterior horn of grey matter. In cases of this kind, the distinction can only be made by observing the mode of development of the symptoms, and the presence or absence of associated symptoms indicative of lead poisoning, such as the blue line on the gums, anæmia, constipation, lead colic, lead rheumatism, etc. In all cases in which a subacute inflammation of the anterior horn of grey matter seems to be present, symptoms and signs indicative of lead impregnation should be diligently looked for. The occupation of the patient is a point of importance; and the possibility of lead being introduced into the system in the water which the patient is in the habit of drinking should be kept in view.

Prognosis.—Undoubted cases of subacute inflammation of the anterior horn of grey matter are so rare that it is difficult perhaps to lay down any definite rules as regards the prognosis.

It would appear, however, that in many cases improvement occurs and that in some the recovery is complete. In the majority of cases in which the paralysis has attained a high degree, some of the affected muscles will probably remain more or less paralysed and atrophied. In some cases, the disease proves fatal; and this result is not unfrequently due, as I have already pointed out, to the extension of the lesion to the medulla oblongata and the development of bulbar symptoms.

Treatment.—The same measures of treatment which are advisable in cases of poliomyelitis anterior acuta, after the febrile stage has subsided, and in cases of progressive muscular atrophy (the treatment of which will be considered in a future lecture), should be adopted.

LECTURE VIII

PRIMARY SCLEROSIS OF THE CROSSED PYRAMIDAL TRACT

GENTLEMEN, if I were to adopt a strictly systematic plan of description, I should now take up progressive muscular atrophy, for the pathological substratum of that disease is a chronic lesion of the anterior horn of grey matter—the region of the cord that we have just been considering. But since progressive muscular atrophy is very often associated with, or complicated by, sclerosis of the crossed pyramidal tract, it will, I think, be more practical and useful to consider in the next place the system disease in which the crossed pyramidal tracts are sclerosed. After you are familiar with the symptoms which result from sclerosis of the crossed pyramidal tracts, you will be better able to understand the symptomatology of progressive muscular atrophy, and especially of amyotrophic lateral sclerosis.

Synonyms.—The term *Primary Sclerosis of the Crossed Pyramidal Tracts* is, I think, better than the terms *Lateral Sclerosis*, *Primary Lateral Sclerosis*, and *Tabes Dorsalis Spasmodique*, which have also been applied to this disease.

The lateral column of the cord contains several tracts of fibres, viz.:—(1) The crossed pyramidal tract; (2) The direct cerebellar tract; (3) The antero-lateral ascending tract and (4) The antero-lateral descending tract (Gowers' tracts); (5) The internal boundary layer of the grey substance; and (6) Lissauer's tract. (See fig. 38.)

The terms lateral sclerosis and primary lateral sclerosis are indefinite—strictly speaking, they imply or ought to imply sclerosis of all of the tracts of which the lateral white column of the cord is composed. The term primary sclerosis of the crossed pyramidal tracts, which is quite definite, is therefore preferable.

The crossed pyramidal tract is a very important tract of fibres. It is the chief continuation in the cord of the great motor or pyramidal tract, which passes down, you will remember, from the motor centres in the cerebral cortex through the corona radiata, internal capsule, pons Varolii, and medulla oblongata, to the lower end of the spinal cord.

At the lower end of the medulla oblongata, the great pyramidal tract divides, as you know, into two bundles of fibres.

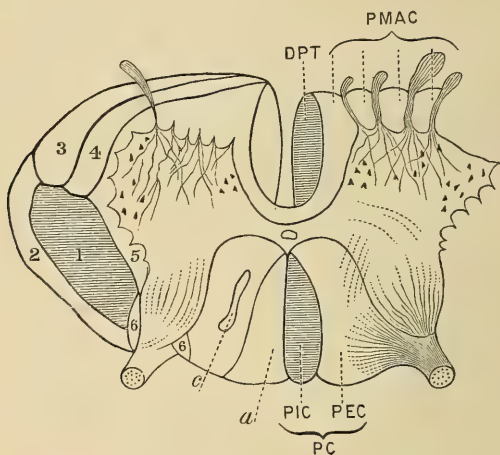


FIG. 38.—*Diagrammatic Section of a Spinal Segment showing the Physiological division of the White Matter.*

The lateral column of the right half of the segment has been cut away. PIC—Postero-internal column. PEC—Postero-external column. DPT—Direct pyramidal tract. PMAC—Principal mass of the anterior column. 1. Crossed pyramidal tract. 2. Direct cerebellar tract. 3. Antero-lateral ascending tract (Gowers' tract). 4. Antero-lateral descending tract. 5. Internal boundary layer of the grey substance. 6., 6—Lissauer's tract. *a*—Postero-internal column. *c*—Comma-shaped tract.

One, the larger bundle, decussates and enters the lateral column of the cord on the opposite side, through which it descends; it is termed the *crossed pyramidal tract*; the other, which is a much smaller bundle of fibres, does not decussate, but passes down on the same side of the cord in the anterior column; it is termed the *direct pyramidal tract*. Further, recent researches seem to show that some of the fibres of the main pyramidal tract in the medulla oblongata pass down the spinal cord (undecussated) in the crossed pyramidal tract of the same side.

As the pyramidal tracts (crossed and direct) descend through the cord, fibres pass off from them and enter the grey matter of the anterior horn. After entering the grey matter, these fibres divide and subdivide and form a leash, as it were, of very delicate fibrils. Until recently, it was supposed that the fine fibrils which are the terminations of the fibres of the crossed pyramidal tracts were directly continuous in the grey matter with the fine fibrils in which the branches of the multipolar nerve cells (other than the axis-cylinder process) terminate. But recent researches seem to show that both sets of fibrils terminate in rounded or bulbed ends; and that although in the closest apposition, the two sets of fibrils are not *directly* and *structurally* continuous with one another.

But be this as it may, voluntary motor impulses which are destined for the limb and trunk muscles, after being discharged by the cortical centres in the brain, pass right down through the pyramidal tract until they reach the anterior horn of grey matter in the spinal cord. They then pass on to the multipolar nerve cells, and thence (through the axis-cylinder processes and motor nerve fibres which are direct continuations of the axis-cylinder processes) to the muscles. (See fig. 39.)

The main function of the crossed pyramidal tract is, therefore, to conduct voluntary motor impulses from the cortical centres, above, to the multipolar nerve cells in the anterior horn of the spinal cord below, and thence to the muscles.

The direct pyramidal tract serves a similar purpose.

The crossed pyramidal tract chiefly conducts motor impulses from the cortical centres on the opposite (say the right) side of the brain to the muscles on its own (the left) side of the body. Further, you must remember (as I have already pointed out) that the crossed pyramidal tract contains some undecussated fibres, i.e. some fibres which conduct voluntary motor impulses from the cortical motor centres on the same side of the brain.

Since it is now believed that the terminal fibres of the crossed pyramidal tracts are not directly continuous with the terminal fibres of the nerve cells, we must suppose that a nerve vibration, concerned in the production of a voluntary movement, produces a corresponding nerve vibration or functional change in the terminal fibres of the multipolar nerve cells. This change may perhaps be compared to the change which a magnet pro-

duces in a bar of soft iron with which it is brought into close juxtaposition, but with which it is not in direct contact.

The exact destination of the motor impulses which are con-

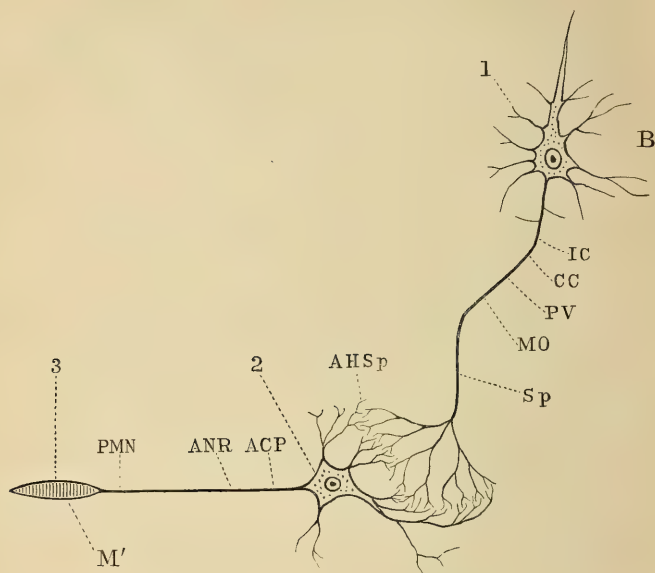


FIG. 39.—*Plan of the motor nerve apparatus.*

Figure 1 points to a large pyramidal nerve cell, in the motor area of the right hemisphere of the brain (B), from which a basal axis-cylinder process passes, as a fibre of the main pyramidal tract, down to the spinal cord, through the internal capsule (IC), crus cerebri (cc), pons Varolii (pV), medulla oblongata (Mo,) and spinal cord (sp). In the grey matter of the spinal cord, this fibre breaks up into a brush of fine fibrils, which are said to terminate in rounded ends.

Figure 2 points to a multipolar (motor) nerve cell in the grey matter of the anterior horn of the spinal cord (AH sp). From this cell an unbranched axis-cylinder process (acp) passes, through the anterior nerve root (anr), peripheral motor nerve (pmn), down to the muscle (M) to which the figure 3 points.

The branched fibres of the multipolar nerve cell in the spinal cord terminate in a brush of fine fibrils, which are supposed to terminate in rounded or bulbed ends, and which are in close apposition with the terminal fibrils of the fibres of the pyramidal tract.

ducted downwards by the direct pyramidal tract (whether to muscles on its own or the opposite side of the body) is still undetermined.

One theory supposes that the motor impulses which are conducted downwards by the fibres of the direct pyramidal tract

pass to nerve cells on the same side of the cord. Another supposes that the fibres of the direct pyramidal tract decussate at their point of termination in the spinal cord, and that the motor impulses which they conduct pass to muscles on the opposite side of the body, i.e. the opposite side to the cortical centres from which they are discharged.

The direct pyramidal tract is not only smaller, but it is also shorter than the crossed pyramidal tract. It usually terminates in the mid-dorsal region of the cord; the fibres which it contains are probably, therefore, for the most part connected with the motor mechanisms (multipolar nerve cells) of the cervical region of the cord (i.e. with the muscles of the upper extremities).

In addition to conducting voluntary motor impulses, it would appear that the fibres of the crossed pyramidal tract conduct impulses which regulate or control reflex movements. I will return to this point, which is of great clinical importance, presently.

The trophic integrity of the fibres of the crossed pyramidal tract depends upon the healthy functional activity of the large pyramidal (motor) nerve cells in the motor cortex of the brain. We have already seen that the nutritive condition of the axis-cylinder process, the motor nerve fibre with which it is continuous, the motor nerve-endings, and muscular fibres in which the motor nerve-endings terminate, depends upon the healthy activity of the multipolar nerve cells in the anterior horn of the spinal cord. What I want now to impress upon you is that the nutritive condition of the long tract of fibres which we term the pyramidal tract, depends upon the healthy condition of the large pyramidal nerve cells in the cerebral cortex.

The neuro-motor nerve tract consists, as we have seen, of two divisions:—(1) An upper division which extends from the motor cortical centres above to the terminations of the fibres of the pyramidal tract below; and (2) A lower division which extends from the multipolar nerve cells in the spinal cord above to the muscular fibres below.

In each of these divisions the branching fibres of the respective nerve cells, which are the trophic centres of the upper and lower divisions respectively, should be included. (See fig. 39.)

The pyramidal nerve cells in the cerebral cortex are the trophic centres for the upper division; the multipolar nerve

cells in the anterior cornua of the spinal cord are the trophic centres for the lower division. Destruction of the nutritive or trophic centre (the large pyramidal cells) in the cerebral cortex leads to the production of secondary descending or nutritive degeneration in the upper division; just as destruction of the multipolar nerve cells in the spinal cord leads to the production of secondary descending or nutritive degeneration in the lower division.

The degeneration of the fibre of the pyramidal tract, which results from destruction of its trophic centre (the pyramidal nerve cell from which the fibre springs) in the cerebral cortex, involves, you will observe, the whole length of the fibre including the fine fibrils in the grey matter of the spinal cord in which the fibre terminates. The degeneration (usually) stops short abruptly at the bulbed terminations of those fibrils. It was difficult to explain this abrupt arrest of the degeneration, so long as it was supposed that there was a direct structural connection between the terminal fibrils of the pyramidal tract and the terminal fibrils of the nerve cells. It is easy to explain the abrupt arrest of the degeneration now that we know (perhaps I should say believe) that the two sets of fibrils, though in close contact, are not directly continuous with one another.

Pathological Physiology.—Let us now pass to the pathological physiology of this important crossed pyramidal tract.

Lesions which interrupt the conduction of motor impulses through the crossed pyramidal tract, of necessity cause loss of voluntary motor power (i.e. paralysis) in the motor mechanisms (muscles) with which it is connected.

The extent and distribution of the paralysis depends upon the height or position of the lesion. A lesion which completely destroys the crossed pyramidal tract as it passes through the internal capsule of the brain, will necessarily produce paralysis in all of the parts (face, arm and leg on the opposite side) which the great motor pyramidal tract innervates. A lesion which completely destroys the fibres of one pyramidal tract in the dorsal region of the spinal cord will produce paralysis of the lower extremity (leg) on the same side; the face and arm muscles will not of course be paralysed.

The extent and distribution, then, of the paralysis which

results from a lesion of the pyramidal tract, depend upon the height or position (in the pyramidal tract) of the lesion. You must remember that at both ends (the upper and lower ends) of the motor apparatus, there is anatomical separation and physiological differentiation. In the cortex of the brain, the fibres of the pyramidal tract are widely separated, for the different cortical centres are spread over an extensive area of the brain surface. In the internal capsule and still more in the crus cerebri, pons Varolii, medulla oblongata, and lateral column of the spinal cord, the fibres of the pyramidal tract are crowded together, as it were, into a narrow area. In the grey matter of the spinal cord and in the parts of the motor tract below the spinal cord, the anatomical separation is even more marked than in the brain, and it reaches its highest degree of separation in the muscles. Hence it follows that the distribution of the paralyses which result from lesions of the pyramidal tract (in the brain, pons Varolii, medulla oblongata, and spinal cord) is very different and variable.

This is a general statement to which I shall frequently refer in subsequent parts of the course.

At present, we are concerned with lesions of the crossed pyramidal tract in the spinal cord; lesions of the direct pyramidal tract may, for our present purpose, be ignored, for they are far less important than lesions of the crossed pyramidal tract.

Lesions, then, which interrupt the continuity or prevent the conduction through the fibres of the crossed pyramidal tract in the spinal cord, produce as their *first effect* loss of motor power (paralysis), the extent and distribution of which depends upon the position, that is to say, the height of the lesion in the cord. A lesion in the cervical region will produce paralysis both of the arm and of the leg on the same side; a lesion in the dorsal region will produce paralysis of the leg on the same side, only, the arm of course escaping. Further, the distribution of the paralysis will of course depend upon the fact whether the lesion is bilateral or unilateral. Now, the spinal cord is, comparatively speaking, a narrow structure. In the spinal cord, the two crossed pyramidal tracts are brought into close juxtaposition; hence it follows that many lesions which destroy the structure of the cord will necessarily implicate the pyramidal

tracts on both sides, that is to say, will produce paralysis on both sides of the body. The paralysis which results from an indiscriminate lesion, say a myelitis, is in the great majority of cases bilateral, i.e. paraplegic in distribution.

The lesion which is the pathological substratum of *the* system disease which we are now considering—*primary* sclerosis of the crossed pyramidal tract—is almost invariably bilateral (see fig. 40); hence the resulting paralysis is also paraplegic in distribution.

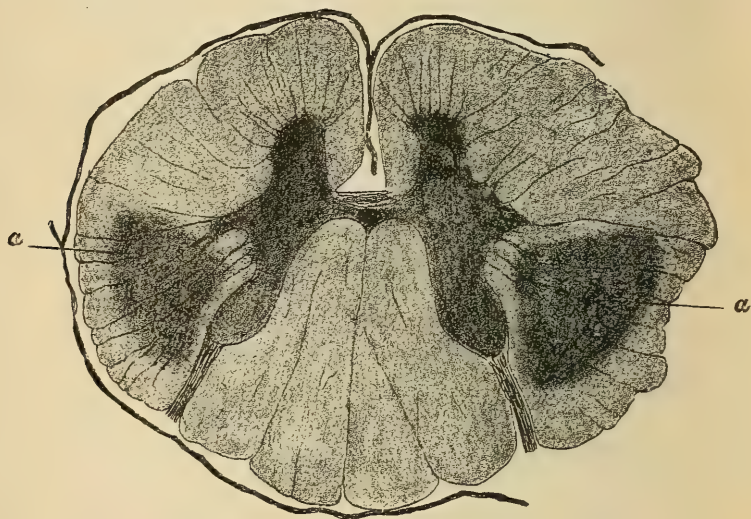


FIG. 40.—*Transverse section through the upper dorsal region of the cord in a case of Primary Lateral Sclerosis. Stained with carmine, mounted in dammar, and magnified about 10 diameters.*

a, a—Sclerosed patches mapping out the region of the crossed pyramidal tracts.

The preparation was made from a portion of cord which was kindly sent me by Dr. Dreschfeld of Manchester.

Further, the extent and completeness of a paralysis which results from a lesion of the crossed pyramidal tract will also depend upon:—(a) the severity of the lesion, (the number of the fibres of the tract which are interfered with and the completeness of the interference or destruction); and (b) the rapidity (acuteness) with which the lesion is established.

In some cases the lesion is so slight that only some of the fibres in the crossed pyramidal tract at the seat of the lesion are

implicated; in other cases all of the fibres at the point of lesion are destroyed.

In some cases, the lesion is acute, as for example, in cases of transverse myelitis; in other cases the lesion is very slowly developed, as for example, in the primary form of sclerosis which we are at present considering. In other cases, again, the lesion is neither very quickly nor very slowly established, as for instance in some cases of Pott's disease of the vertebræ, in which pressure on the cord produces as its chief effect impaired or arrested conduction through the fibres of the crossed pyramidal tract.

When I come to describe acute transverse myelitis, we shall see that the paralysis (like the paralysis in cases of poliomyelitis anterior acuta) may be partly due to temporary and removable causes (shock, inflammatory œdema and pressure) and partly due to permanent destruction.

Let me sum up the more important points connected with the physiology and pathological physiology of the great motor (pyramidal) tract. It is the channel through which voluntary motor impulses pass from the brain above to the muscles below. Lesions which destroy the fibres of the pyramidal tract or interrupt conduction through them necessarily produce paralysis. Further, in order to intelligently understand the effects which lesions of this important tract produce, it is essential to remember that the nerve fibres of which it is composed are nourished by the large pyramidal motor nerve cells in the cortex of the brain; and that lesions which destroy these trophic nerve cells produce a process of secondary or nutritive descending degeneration, which passes down from the seat of the lesion to the terminations of the pyramidal fibres.

You must also remember that lesions which shut off the trophic influence of the trophic centres necessarily produce secondary or nutritive degeneration in the fibres of the pyramidal tract below the lesion. For example, a lesion which destroys the crossed pyramidal tract in the lower dorsal region of the cord will be followed by secondary descending or nutritive degeneration of the fibres of the pyramidal tract in the lumbar and sacral portions of the cord. The fibres of the pyramidal tract above the lesion will not of course exhibit any degenerative change, for they still receive the trophic influence of the nerve cells in the cerebral cortex.

These facts are of great importance. The degeneration stops abruptly, you will observe, at the end-terminations of the pyramidal tract; it does not extend to the lower (spinal) trophic centre. This explains the very different condition which the paralysed muscles present when the lesion involves the upper and lower divisions of the neuro-motor apparatus respectively.

In our study of poliomyelitis anterior acuta, we have seen that a lesion which destroys the multipolar nerve cells of the anterior horn of grey matter (the lower or spinal trophic centre) or which interrupts the trophic influence, which under normal circumstances the multipolar nerve cells exert upon the muscles, produces not only paralysis, but marked atrophy of the paralysed muscles, abolition of their reflex movements, and, if the lesion is acute, the reaction of degeneration.

A lesion which is situated above the lower (spinal) trophic nerve centre, in any part of the upper segment of the neuro-motor nerve apparatus, also produces paralysis, but the condition of the paralysed muscles is quite different. They do not become markedly and rapidly atrophied; they do not present the reaction of degeneration and their reflexes are not abolished; on the contrary their deep reflexes are exaggerated. (In exceptional cases of hemiplegia, the muscles become markedly atrophied; in cases of this kind it is probable that the secondary degeneration does not stop short at the end-terminations of the pyramidal fibres but involves the multipolar nerve cells in the spinal cord.)

Rigidity and tension in the paralysed muscles and exaggeration of the reflexes, particularly of the deep reflexes, are the most important, *the* characteristic features of a paralysis which results from a lesion of the crossed pyramidal tract, just as atrophy, flaccidity and (if the lesion is acute) the reaction of degeneration are *the* characteristic features of a paralysis due to a lesion in the lower part of the neuro-motor nerve apparatus. The rapid form of atrophy is, you will remember, only developed in those cases in which the lesion is acute.

Sclerosis of the crossed pyramidal tract produces, then, as its *first effect* paralysis; and as its *second effect* rigidity and tension of the paralysed muscles and exaggeration of the deep reflexes.

The exact manner in which this exaggeration of the deep reflexes is produced, is a disputed point. Several theories have been advanced to account for it. One of them supposes that the

reflex centres in the spinal cord are controlled or held in check by a higher centre in the brain, and that this controlling influence is effected through the fibres of the crossed pyramidal tract. Lesions of the pyramidal tract interrupt the inhibiting or restraining power of the upper centre; the lower spinal reflex being uncontrolled is free to discharge in an exaggerated manner, with the result that peripheral irritations which in health would produce little or no obvious effect, cause rigidity and tension in the paralysed muscles and marked exaggeration of the deep reflexes. According to this theory, the fibres of the crossed pyramidal tract may be regarded as reins which, under normal circumstances, are held tight by the brain; a lesion of the pyramidal tract abolishes the control and allows the lower spinal reflex centre to have its head; peripheral irritations (which may be likened to a spur or whip) applied under such conditions produce exaggerated effects. That is a simple and beautiful theory, and perhaps the best which we possess, but it is doubtful if it is correct. However beautiful a theory may appear to be, it is of course useless unless it is correct—unless it satisfactorily accounts for all the facts.

Within the last few years, it has been stated that if you completely remove the control of the brain from the reflex centres in the spinal cord, instead of producing exaggeration you produce abolition of the deep reflexes. Dr. Charlton Bastian, who advanced this view, states that when the spinal cord is crushed and completely destroyed in the dorsal region (when for example its whole thickness is absolutely destroyed by a transverse myelitis), the deep reflexes in the lower limbs are completely and permanently abolished, even though the lumbar enlargement is not directly implicated by the lesion. He maintains that when the lesion *completely* severs the connection between the brain and the lower part of the spinal cord, the reflexes in the portions of the cord below the lesion are permanently abolished. With Dr. Hughlings Jackson, he believes that the cerebellum exerts an enforcing influence upon the lower (spinal) reflexes. Dr. Bastian, Dr. Hughlings Jackson and other good observers have published cases in support of this view, which is, I may say, diametrically opposed to the view which was generally accepted only a few years ago.

But whether Dr. Charlton Bastian's view is correct or not, and

I must say that speaking for myself I have not met with any case which supports it, lesions which involve the crossed pyramidal tracts are, in the vast majority of cases, attended with exaggeration of the deep reflexes. It very rarely indeed happens that the cord lesion is so complete as to entirely interrupt all conduction through it. I repeat that the rigidity and tension of the paralysed muscles and exaggeration of the deep reflexes are most important characteristics of this form of paralysis.

Another theory, which has been advanced to explain the exaggeration of the deep reflexes which results from a lesion in the crossed pyramidal tract, supposes that the lesion is attended with irritation, which, extending downwards, causes a condition of hyperexcitability in the reflex centre (i.e. the multipolar nerve cells of the anterior horn). This view seems to me highly probable.

The absence of atrophy and the reaction of degeneration in cases of paralysis due to a lesion of the crossed pyramidal tract is, of course, explained by the fact that the multipolar nerve cells—the trophic nerve centres—are unaffected.

The *positive symptoms*, then, which result from a lesion in the crossed pyramidal tract are:—(1) Loss of voluntary motor power (paralysis) in the motor mechanisms (muscles) with which the fibres which are implicated are connected; (2) Rigidity and tension of the paralysed muscles and exaggeration of their deep reflexes.

While the *negative symptoms* are:—(1) Absence of marked atrophy in the paralysed muscles; (2) Absence of the reaction of degeneration; (3) Absence of any marked affection of the bladder and rectum (paralysis of the sphincters); and (4) Absence of sensory disturbances, for the crossed pyramidal tract is, as we have seen, a motor tract.

The exact extent and distribution of the paralysis and the rapidity with which it is developed vary of course in different cases, and depend upon the position of the lesion and the rapidity with which it is established.

These preliminary considerations will enable you to understand the symptoms which result from disease of the crossed pyramidal tract.

Lesions of the crossed pyramidal tract may be primary or secondary.

Primary sclerosis of the crossed pyramidal tract is a typical system disease. The lesion extends throughout a long extent, often indeed from the top to the bottom of the cord; in some cases, the lesion is not confined to the cord, but extends up into the medulla oblongata, pons Varolii, or even to the internal capsule of the brain itself. The effect of the lesion is to slowly and gradually impair the functional activity of the pyramidal fibres.

The lesion is bilateral and symmetrical. It very rarely happens that the pyramidal tract on one side only is involved; but it occasionally happens that the pyramidal tract on one side is more extensively affected than its fellow on the opposite side.

In the vast majority of cases, the lesion commences in the lower part of the cord. Consequently, the symptoms, which may be shortly described as those characteristic of spastic paraplegia, are in the great majority of cases first developed in the lower extremities.

Primary sclerosis of the crossed pyramidal tracts is, so far as our pathological knowledge enables us to judge, a rare disease. When I say primary sclerosis of the crossed pyramidal tracts is rare, do not misunderstand me. Do not suppose that sclerosis of the crossed pyramidal tracts and spastic paraplegia are rare. They are exceedingly common. But sclerosis of the crossed pyramidal tracts and spastic paraplegia are not synonymous with *primary* sclerosis of the crossed pyramidal tracts and with *primary* spastic paraplegia. Spastic paraplegia is a general clinical term, applied to a group of symptoms (loss of motor power in the lower extremities with tension and rigidity of the paralysed muscles and increase of the deep reflexes), which may be due to a number of different conditions—to any lesion which produces sclerosis, whether primary or secondary, of the pyramidal tracts on both sides of the cord. In the great majority of cases of spastic paraplegia, the sclerosis of the crossed pyramidal tracts is a secondary degeneration. The most common cause of spastic paraplegia is transverse myelitis; it is a frequent result of disseminated (cerebro-spinal) sclerosis, and is sometimes due to syringomyelia. It sometimes results from pressure on the spinal cord (the result of caries of the vertebræ, pachymeningitis, extra medullary tumours, etc.). Bilateral spastic paraplegia is occasionally due to a lesion on the surface of the

brain which involves the motor centres for both lower extremities. The infantile form of spastic paraplegia (which in many cases is nothing more or less than a bilateral hemiplegia) is usually the result of an injury to the brain at the time of birth—an extravasation of blood on the surface of the brain which causes a meningo-encephalitis, and results in the production of non-development, atrophy and sclerosis of the brain cortex. Spastic paraplegia also results from the rare lesion termed porencephalie. The lesion usually involves the centres for the lower extremities, which at the vertex are situated, you will remember, in close juxtaposition on opposite sides of the great longitudinal fissure. The development of the pyramidal tracts may be interfered with, or a sclerosis produced in the fibres of the pyramidal tracts which pass downwards from the leg centres. I shall return to this subject when I come to describe hemiplegia and the cerebral lesions with which hemiplegia is associated.

Sclerosis of the crossed pyramidal tracts is not unfrequently associated with sclerosis and degeneration of other tracts or areas of the spinal cord. In progressive muscular atrophy, some sclerosis of the crossed pyramidal tracts is often present. In some cases in which the posterior columns of the cord are sclerosed and degenerated, the crossed pyramidal tracts are also affected; the term postero-lateral sclerosis is usually applied to this condition.

From these statements you will perceive that it is necessary to draw a distinction between sclerosis and primary sclerosis of the crossed pyramidal tracts—between spastic paraplegia and primary spastic paraplegia.

In the common form of spastic paraplegia which results from transverse myelitis, sensory symptoms and derangements of the bladder and rectum are almost always present in addition to the motor symptoms—paraplegic spastic symptoms. But in primary spastic paraplegia there are no marked sensory symptoms, and the functions of the bladder and rectum are not usually deranged, at all events in any marked degree. I shall return to these points when I come to speak of transverse myelitis.

I repeat that in primary sclerosis of the crossed pyramidal tracts, which clinically seems to be common enough, but which pathologically (*really*) appears to be a very rare lesion, the inflammatory process is very slowly and gradually established; and

that in the vast majority of cases it seems to have its starting-point in the lower part of the cord. It is usually supposed that the sclerotic process begins in the nerve elements (the nerve tubes of the crossed pyramidal tracts); but in some cases the connective tissue which surrounds the nerve tubes is perhaps first affected. But be this as it may, the result is degeneration, atrophy and destruction of the nerve tubes, and increase of the connective tissue in which the nerve tubes are embedded and by which they are surrounded. In old standing cases, the connective tissue trabeculae throughout the lateral columns, and indeed throughout the whole transverse section of the cord, may be increased in density and thickness—in other words, a tendency to diffuse sclerosis may be present. But in the early stages of typical cases, the lesion is, so far as we know, limited to the region of the crossed pyramidal tracts.

Etiology.—As yet we are ignorant of the exact cause of the lesion, of the inflammatory process which produces the sclerosis.

The facts that the lesion is, in its early stages, limited to the crossed pyramidal tracts, and that the sclerosis begins in the lower end of the cord, perhaps suggest that the pathological change is due to defective (nutritive) action of the cortical centres. We have seen that the fibres of the pyramidal tract, which are nourished from the cortex, are of great length. It is not difficult therefore to understand that, if the cortical nutritive centres are functioning imperfectly, the parts which are farthest removed from the trophic centre, i.e. the terminations of the pyramidal fibres, will be first affected. If this view is correct, the lesion may in some degree be regarded as a form of secondary degeneration.¹

The comparative difficulty with which the blood supply in the lower part of the cord (lumbar enlargement) is maintained affords, I think, the most probable explanation of the fact that the lesion begins in the lower part of the cord. The healthy nutritive condition of the fibres of the pyramidal tract necessarily depends in some degree upon the condition of the vascular blood

¹ The fibres of the pyramidal tract which terminate in the lumbar enlargement of the cord are somewhat larger than those which terminate in the cervical enlargement. This is perhaps a possible, though I do not think a probable, explanation of the fact that the lesion begins in the lumbar enlargement.

supply. We may suppose, then, that a double cause of defective nutrition in the pyramidal fibres of the lumbar region of the cord is present—viz., (1) Defective trophic nerve force, the result of impaired functional activity of the pyramidal cells of the motor cortex) which favours the production of, or tends to produce, degeneration *in the terminal fibres* of the pyramidal tract in the lumbar and cervical regions of the cord); and (2) Defective blood supply in the lower end of the cord (which determines that the defective nutrition in the terminal fibres of the pyramidal tracts should be *first* manifested at the lower end of the cord, i.e. in the lumbar enlargement).

Although the sclerosis usually begins in the lower end of the cord, it gradually extends upwards as the disease progresses, and it may ultimately involve the fibres of the crossed pyramidal tracts in the cervical enlargement. In other words, the sclerosis seems to extend in a direction opposite to the direction in which the affected nerve fibres functionate. Now, as a rule, degenerative processes extend along nerve fibres in the direction of their functional activity. But it is certain that nerve tubes can conduct in two directions; and it cannot be denied that a degenerative process may, and undoubtedly in some instances does, pass in a direction which is opposed to the usual direction in which the nerve tubes functionate (i.e. conduct).

From these statements you will see that we have still much to learn regarding the pathology and etiology of this lesion.

It is highly probable, I think, as Dr. Gowers supposes, that in some cases the lesion commences in the grey matter of the cord, i.e. in the terminal nerve fibrils of the crossed pyramidal tracts. He suggests that the sclerosis is in some cases the result of a toxic irritant which has a special affinity for the terminal fibres of the pyramidal tract (the terminal fibres of the upper segment of the neuro-motor apparatus), just as alcohol and some other poisons seem to have a special affinity for the terminal fibres of the lower segment of the neuro-motor apparatus (i.e. the terminal fibres of the peripheral nerves).

It is not improbable that sclerotic and inflammatory lesions, limited to the crossed pyramidal tract or the terminal fibres of the tract, may be due to several different causes.

Primary sclerosis of the crossed pyramidal tracts is essentially a disease of the adult; in most cases it commences between

the ages of thirty and forty-five, but it may develop earlier or later than this. It is practically speaking unknown in childhood; care must be taken not to confound it with the infantile form of spastic paraplegia which is, as we have seen, a totally different condition. The disease seems to be more common in men than in women, although cases which present all the clinical characters of the disease are not unfrequently seen in young women between the ages of sixteen and twenty-five or thirty. Some of the cases which at their commencement are apparently cases of primary spastic paraplegia, ultimately turn out to be cases of multiple or disseminated sclerosis. I have met with several cases of this kind in young and middle-aged women. I shall refer to this point in more detail when I come to speak of cerebro-spinal sclerosis.

Exposure to cold, traumatic injury, and overuse of the affected muscles have been thought in some cases to be the exciting cause of the lesion. Syphilis has also been blamed, but it is very doubtful if it is a cause of any importance. Of course, in some cases of primary sclerosis of the crossed pyramidal tracts, a history of previous syphilis can be elicited; but the same statement may be applied to every known disease. A history of previous syphilis in a few individual cases is not of much importance; and almost all observers seem to be agreed that it is only in a comparatively small proportion of cases of primary spastic paraplegia that a history of syphilis is forthcoming. In this respect, primary spastic paraplegia contrasts remarkably with locomotor ataxia.

I do not of course mean to say that the clinical groups of symptoms comprised under the term spastic paraplegia are seldom the result of syphilis, for as Erb and others have pointed out, and as every one with a large clinical experience knows, spastic paraplegia is the usual result of syphilitic and gummatous myelitis. What I do say is that *primary* spastic paraplegia is, so far as we know, very rarely indeed the result of syphilis.

In a few cases, hereditary influences seem to play a part in the production of the disease; but this, too, is rare.

LECTURE IX

PRIMARY SCLEROSIS OF THE CROSSED PYRAMIDAL TRACTS (*Continued*)

Clinical History.—Let us now turn to the clinical features. Primary sclerosis of the crossed pyramidal tract is a very chronic disease, in fact it is probably *the* most chronic disease of the spinal cord with which we are at present acquainted. The onset is usually slow and gradual. In the vast majority of cases, the symptoms commence, as we have seen, in the lower extremities, sometimes in one lower extremity. The early symptoms are weakness, heaviness, and stiffness in the legs. As the disease progresses the symptoms become more marked and the difficulty in walking becomes greater, until finally a condition of complete spastic paraplegia is developed. The stiffness, which is usually most marked the first thing in the morning when the patient begins to walk, in many cases lessens after exercise, just as the stiffness in a 'groggy' horse often passes off after a little exercise. The difficulty in walking, stiffness and rigidity are usually increased by exposure to cold, and lessened by warmth. External irritation of any kind is apt to increase the muscular rigidity and tension; it is probably in consequence of this fact that patients affected with spastic paraplegia usually walk better in their shoes and stockings than when bare-footed.

As the disease progresses, walking becomes more and more difficult. In fully developed cases, the gait is highly characteristic. The patient usually walks with one or two sticks. Each step is made with difficulty; the feet appear to be stuck to the ground, or rather the foot which is about to be brought forward appears to be stuck to the ground. In order to raise it from the ground, a powerful effort has to be made with the muscles of the trunk and upper part of the body. There is little or no flexion of the hip; the pelvis and lower limb are raised as

a whole; the patient leans heavily on his stick; the back is arched; the chest is thrown forward. As the leg is advanced, the toes scrape against the ground with an unpleasant grating noise; the knees tend to interlock as the foot comes forward; at the end of the step, the foot often comes forward with a jerk and tends to cross its fellow on the opposite side.

The gait of spastic paraplegia is pathognomonic; you can recognise the condition at the first glance. The gait is very different from that of locomotor ataxia, which, as we shall afterwards see, is also highly characteristic.

In other cases, when the leg is raised from the ground, a peculiar hopping movement of the body occurs, which Erb thinks is due to spasmodic contraction of the calf muscles. If the patient should happen to rest his weight on the balls of his toes (in going up or down stairs or when sitting in a chair, for example) a rhythmical clonic tremor (a spontaneous ankle-clonus or *trépidation spontanée*, as the French term it,) is often produced.

The muscles are firm, hard, well-developed and well-nourished; in some cases they even appear to be hypertrophied, and in some cases there is perhaps an actual increase in the size of the muscular fibres. But although the muscles are well nourished some evidence of motor impairment can generally be detected on careful examination; it is often most marked in the flexors of the ankle; the extensors of the knee, on the contrary, are usually very powerful; in many cases, one is unable to bend the leg, if the patient chooses to keep it straight.

The absence of wasting and the presence of rigidity and tension in the affected muscles are very characteristic features.

In pure, typical and uncomplicated cases of primary spastic paraplegia, there is no muscular atrophy during the earlier stages of the case. But it must be remembered that in many of the cases in which the sclerosis of the crossed pyramidal tract appears to be primary, some muscle or perhaps some groups of muscles, are atrophied; for the sclerotic process in the crossed pyramidal tract is in some cases associated with degeneration of the anterior horn of grey matter.

As I have already remarked, it is only in quite exceptional cases that the sclerosis of the crossed pyramidal tract is limited to one side of the cord, but the associated degeneration of the

anterior horn is often limited to one side, or at all events much more marked on one side than on the other.

Exaggeration of the reflexes and especially of the deep reflexes, is another highly characteristic feature of spastic paraplegia—whether primary or secondary. The knee-jerks are greatly increased and ankle-clonus can almost always be elicited; in some cases, a knee-clonus can be obtained; exceptionally, the knee-jerk is excessive, but there is no ankle-clonus; I have seen several cases in which a knee-clonus was present, but in which there was no ankle-clonus; this is, however, rare. These modifications depend, of course, upon the distribution of the sclerosis in the cord, and the exact manner in which the individual muscles are affected.

The increase of the deep reflexes may still be detected in those cases in which the sclerosis of the crossed pyramidal tracts is complicated by degeneration of the anterior horns of grey matter, unless indeed the whole of the nerve cells in the anterior horns should happen to be destroyed.

The superficial, like the deep, reflexes, are usually exaggerated. But it is not always easy to demonstrate the increase; for, in advanced stages of the disease, when the legs are rigidly extended like iron bars, tickling the soles may not produce any definite reflex movement; it may merely cause a greater rigidity of the already rigid limbs. Nevertheless, in many cases, marked exaggeration of the plantar reflex is a very striking feature. In some cases, the whole limb is thrown into a condition of clonic spasm—spinal epilepsy—by gently tickling the sole of the foot.

In this form of paralysis, the electric condition of the affected muscles is rarely altered in any marked degree. During the early stages of the disease, the electrical excitability to both forms of current appears in some cases to be increased (this change is termed a 'simple increase'). As the disease progresses, the excitability to both forms of current usually diminishes (this change is termed a 'simple diminution'). In typical and uncomplicated cases, the electrical reactions do not present any *qualitative* changes throughout the whole course of the disease; in other words, the reaction of degeneration is never developed.

The sensory functions are, in most cases, quite normal. In some cases, it is true, the patient complains of aching pains in the back, or rather of muscular weariness than of actual pain. In

some cases, too, muscular pains, often due to spasm, are felt in the legs. But in typical cases, the sensory functions of the skin are unaffected. Patients not unfrequently complain of numb-

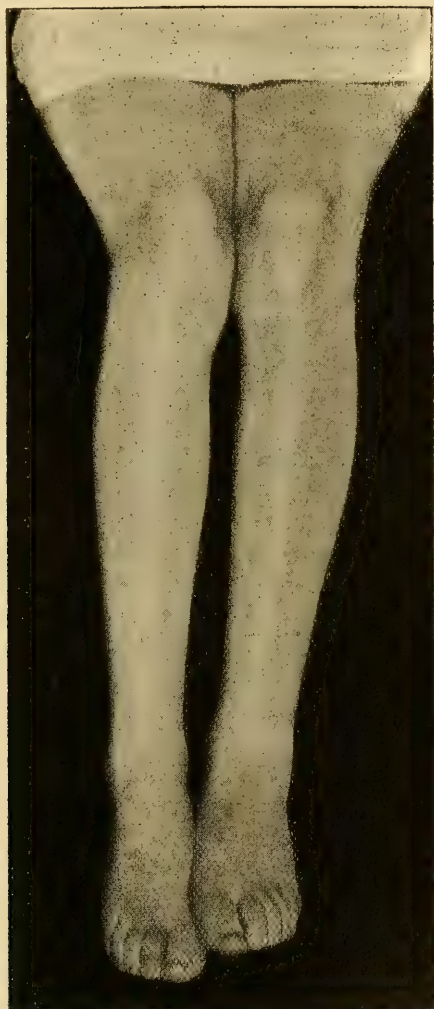


FIG. 41.—*Spastic paraplegia, second stage; the legs are rigidly extended and adducted.*

ness, tingling, or a feeling of 'deadness' as they term it, in the legs; but there are no definite and distinct *objective* signs of sensory impairment, no definite anæsthesia, unless of course the sclerosis in the crossed pyramidal tract is complicated with sclerosis of the posterior column.

In typical cases, the functions of the bladder and rectum are not affected in any marked degree—the sphincters are rarely paralysed, even during the later stages of the case. The expulsive power of the bladder is in some cases impaired. In other cases, the condition which I term precipitate urination is present. It seems to depend upon diminution or abolition of the voluntary control which the brain normally exercises over the vesical centre in the cord, i.e. over the act of micturition. The result is that when the bladder becomes distended the discharge of the vesical centre cannot be restrained by the inhibitory power of the will; when the desire to urinate is felt, the patient is obliged to make water there and then. In women, some difficulty in urination may be caused by the inability to separate (abduct) the rigid legs.

Constipation is common; but unless it is very marked, one should hesitate to attribute it directly to the cord lesion. Constipation may of course be due to many different conditions; and in this disease, in which the power of walking is so much impaired, it is often due to want of muscular exercise. In many cases of primary spastic paraplegia, constipation is not a marked symptom; the reflex irritation and increased excitability of the reflex centres in the lower part of the cord prevent this.

Optic atrophy, which is not uncommon in sclerosis of the posterior columns (locomotor ataxia) is, practically speaking, never met with in primary sclerosis of the crossed pyramidal tracts. Gowers states that nystagmus may often be detected. I have only very rarely observed it, perhaps because I have not specially looked for it; and I am disposed to think that in many of the cases in which nystagmus is present, the spastic paraplegia is probably due to commencing disseminated sclerosis rather than to primary sclerosis of the crossed pyramidal tracts. The same statement may perhaps be applied to the rare cases of primary spastic paraplegia in which optic atrophy is developed.

Up to this point I have been speaking of the condition of

the lower extremities, in which the symptoms of the disease are chiefly located; but you must remember that it not unfrequently happens that the upper extremities are affected in some degree. Patients affected with primary spastic paraplegia not unfrequently complain of subjective sensations, such as numbness, deadness, and 'pins and needles' in the finger tips, and of weakness in the hands and arms; but marked paralysis, rigidity and tension are very rarely developed in the upper limbs or in the face. The muscular or myotatic irritability of the upper limbs is often increased; the triceps-jerk and wrist-jerk are in many cases exaggerated; and a jaw-jerk or jaw-clonus can in some cases be elicited.

In very exceptional cases, the loss of motor power and rigidity are limited to one arm and one leg.

Such are the leading clinical features of the *first stage* of the disease—the stage of *incomplete spastic paraplegia*, during which the patient is able to walk about usually with the aid of a stick or sticks.

As the disease progresses, the difficulty in walking gradually increases until finally the patient may be unable to walk at all. This, the *second stage* or the stage of *complete spastic paraplegia*, as it may be termed, is in some cases never reached, and in most cases is only reached after the first stage has continued for a long time. I have at present under observation a man who suffers from the most marked spastic paraplegia, apparently primary; although the disease has been in existence for thirty years, he is still able to walk about; in his case, the second stage has not as yet been reached.

During the second stage, the patient lies in bed with the legs in a condition of rigid extension. The muscular nutrition may remain good; the muscles may still be firm and tense; the exaggeration of the deep reflexes continues; any external irritation, such as is produced by handling the limbs or tickling the soles, increases the rigidity. During this stage, twitchings and jerkings are apt to occur in the legs. In describing the clinical phenomena of the first stage, I should have told you that twitchings, jerkings, and spasmodic contractions of the affected muscles are of frequent occurrence; in some cases, they chiefly occur during sleep, or just as the patient is going to sleep or waking from sleep.

During the second stage of the disease, the nutrition of the affected muscles is usually well maintained, though a certain amount of wasting is not uncommon. The electrical excitability may present some ('simple') diminution, but in many cases it remains normal, or for practical purposes normal; the sensory functions are usually unaffected and paralysis of the sphincters of the bladder or rectum is of rare occurrence—unless of course, the lesion should extend to and involve other areas of the cord than the crossed pyramidal tracts. During the second stage of the disease, the arms may become affected, or if they happen to have been implicated during the first stage (but this, as we have seen, is rarely the case), the weakness and stiffness in the upper extremities become more marked.

During the second, as during the first, stage of the disease, the general health is usually unimpaired, the digestive apparatus works satisfactorily, the state of nutrition is well preserved, and complications on the part of the heart, lungs and other organs are, comparatively speaking, rarely developed.

The second, like the first, stage may continue for many years. Ultimately the patient dies from some intercurrent complication or disease, or the third stage of the disease is reached.

The *third stage* of the disease is characterised by the extension of the morbid process and the development of complications (such as cystitis, kidney disease, etc.) which can be directly attributed to the cord lesion. It must be remembered that the three stages of the disease run insensibly one into the other; the division into stages, though useful for practical and clinical purposes, is in some degree artificial.

During the third stage, the morbid process may extend to the anterior cornu and lead to the production of muscular atrophy; or to the posterior columns and produce well marked sensory derangements, etc. It must be remembered that this extension may occur at any period of the disease; but it is much more frequent during the later than during the earlier stages. In this disease the risk of grave bladder and urinary complications (cystitis, pyelitis and surgical kidney) is much less than in some of the affections of the spinal cord, such as myelitis and locomotor ataxia, which we shall afterwards have to consider. This is a most important point to which I will refer in more detail in connection with the prognosis. The

reason why grave bladder and urinary complications are, comparatively speaking, so rarely developed in primary sclerosis of the crossed pyramidal tracts seems to be that the paralysis is rarely complete and that the urine does not stagnate and undergo septic changes in the bladder. The increased reflex excitability which is such a prominent feature of the disease, ensures the frequent emptying of the bladder, even in those cases in which the organ is not under proper voluntary control.

I have now described all the more important points connected with the clinical history of primary sclerosis of the crossed pyramidal tracts. Let me repeat that sclerosis of the crossed pyramidal tracts, apparently primary, is frequently met with in association with progressive muscular atrophy, that it is sometimes combined with sclerosis of the posterior columns of the cord, constituting the disease which we term postero-lateral sclerosis; and that it is sometimes also seen in association with general paralysis of the insane. Further, do not forget that in the great majority of cases in which the crossed pyramidal tracts are sclerosed (i.e. in which spastic paraplegia is present), the sclerosis is not primary but secondary, the result of myelitis, cerebro-spinal sclerosis, pressure on the cord, etc. Again, let me repeat that the infantile form of spastic paraplegia is due to a cerebral lesion, the lesion in the cord is merely a secondary nutritive degeneration or an arrested development of the crossed pyramidal tracts.

Diagnosis.—In typical and well marked cases, the diagnosis of spastic paraplegia presents no difficulty. The *first step* in the diagnosis of *primary* spastic paraplegia—the conclusion that the lesion involves the crossed pyramidal tracts—is easily made, and is based upon the presence of motor weakness, stiffness and rigidity, and especially of marked exaggeration of the deep reflexes (the presence of ankle-clonus and great exaggeration of the knee-jerks).

The *second step* in the diagnosis—the conclusion that the lesion is limited to the crossed pyramidal tracts—is attended with more difficulty, for sclerosis of the crossed pyramidal tracts is not synonymous with *primary* spastic paraplegia. In order to attain the second step in the diagnosis, it is necessary to exclude several other conditions, such as transverse myelitis, cerebro-spinal

sclerosis and Pott's disease of the vertebræ, in which secondary descending degeneration and (secondary) spastic paraplegia are frequently developed. Further, it is of course necessary to exclude progressive muscular atrophy, postero-lateral sclerosis, and the infantile form of spastic paraplegia which is, as we have seen, the result of a cerebral lesion.

The recognition, then, of primary sclerosis of the crossed pyramidal tracts is based, *firstly*, on the presence of certain *positive* symptoms (muscular weakness, tension, rigidity and exaggeration of the deep reflexes) which show that there is a lesion of the crossed pyramidal tracts; and *secondly*, on the absence of certain other *negative* symptoms (such as anæsthesia, muscular atrophy, inco-ordination, etc.) which show that the lesion is confined to the crossed pyramidal tracts (i.e. that the other areas and tracts of the cord are not implicated). The presence, if the term may be allowed, of these two sets of symptoms (positive and negative) justifies the conclusion that the cord lesion is limited to the crossed pyramidal tracts.

The *third step* in the diagnosis, viz., the conclusion that the disease is the system disease—primary sclerosis of the crossed pyramidal tracts—is warranted if (*a*) the disease has developed slowly in an adult (thereby excluding sclerosis in the crossed pyramidal tracts due to a lesion of the cortical centres or in early childhood or at the time of birth); and (*b*) provided that the condition (the spastic paraplegia) is not hysterical.

This reasoning may perhaps seem somewhat detailed, and as regards the last step, perhaps somewhat too guarded and qualified; but the reasoning is not difficult to follow and the detail and qualifications are necessary.

The first step in the diagnosis is to determine that there is a lesion in the crossed pyramidal tracts; the second to show that the lesion is limited to the crossed pyramidal tracts; and the third to show that the lesion is slowly developed, is organic, and is primary, in other words, that it is *the particular* sclerotic lesion which is the pathological substratum of *primary* spastic paraplegia.

The differential diagnosis of primary sclerosis of the crossed pyramidal tracts and of hysteria.—I have said that it is necessary to exclude hysteria, and this leads me to say that in its early stages primary spastic paraplegia is very liable to be mistaken for

hysteria. Mistakes of this kind frequently occur. Hysteria may imitate every organic disease of the nervous system; and it has been well said that the imitation is in many cases 'so exact as to deceive the very elect.' Primary sclerosis of the lateral columns is no exception to this statement. In this country, hysterical paraplegia is rarely met with in the male, it is essentially a disease of young women. But the mere age and sex of the patient are not conclusive, for primary spastic paraplegia, if we may rely upon the clinical evidence alone, is not very uncommon in young and middle-aged women. Ignorance of these facts is one of the chief reasons why the two diseases are so frequently confounded. Another reason is, that in primary spastic paraplegia, the nutrition of the muscles is well maintained. Some people seem to think that if the muscles are well developed, paralysis due to organic disease is not likely to be present. It is unnecessary to say that such a conclusion is a most mistaken one.

Further, the presence of other hysterical symptoms does not conclusively show that the paralysis is functional (i.e. hysterical). The presence of hysterical symptoms certainly shows that the patient is suffering from hysteria, but hysteria and organic disease are often combined. The presence of hysterical symptoms does not exclude organic disease—does not show that the *only* thing from which the patient is suffering is hysteria. These are very important statements which I shall repeat again and again throughout the course. Let me beg of you to remember that you are never justified in coming to the conclusion that the symptoms are *merely hysterical*, unless you are satisfied, after a careful and detailed examination of the whole nervous system, that *there are no symptoms and signs indicative of organic disease*. Let me repeat that organic disease is very often associated with hysteria, and is frequently the cause of the hysterical condition. When we come to study hysteria, I shall have to tell you that anything (traumatic injury or disease in the pelvic viscera, for example), which lowers the nerve tone and disturbs the nerve health, is very apt, particularly in women, to produce the condition which we term hysteria. You will now readily understand why it is that hysterical symptoms are so frequently associated with organic disease of the nervous system, and especially with organic lesions of the brain, which lower the

nerve tone and the nerve control—in other words, which produce the physical basis, so to speak, of the hysterical condition.

The age and sex of the patient and the fact that the patient is certainly hysterical are not, therefore, of themselves conclusive. Of course the presumption is that a paralysis in a young hysterical woman is functional. Under such circumstances, we naturally first think of hysteria as a cause for the paralysis. But a presumption is not a diagnosis. If we wish to arrive at a satisfactory conclusion—to make a diagnosis—we must go further than this.

The exact distribution of the paralysis, the condition of the paralysed muscles, the condition of the reflexes, the condition of the bladder and rectum, and the condition of the sensory functions must be carefully investigated and taken into account.

Hysterical paralysis is much more frequently hemiplegic than paraplegic in distribution, and hemiplegia due to primary sclerosis of the crossed pyramidal tract is, as we have seen, a very rare condition. It is only, therefore, in the, comparatively speaking, rare cases in which the hysterical paralysis is paraplegic in distribution that primary spastic paraplegia is simulated. In hysterical paralysis, the muscles are usually flaccid; but this is not always so, for marked rigidity and contracture are sometimes present. But in those cases of hysterical paraplegia in which the muscles are rigid, the spasmodic contraction is not identical with that of primary spastic paraplegia. Gowers lays particular stress upon the peculiar extensor character of the spasm, lessening with flexion, which is characteristic of spastic paraplegia due to organic disease; nothing, he says, resembling this ever occurs in hysteria. I can hardly go so far as this, for I am prepared to meet with almost anything in hysteria; but the peculiar character of the spasm is undoubtedly of considerable diagnostic significance.

In hysterical paraplegia, the knee-jerk reflexes are often exaggerated, but typical ankle-clonus can very rarely be elicited. There are perhaps some exceptions even to this statement; and in hysterical cases an imperfectly developed form of ankle-clonus is occasionally met with.

In hysterical paraplegia, symptoms indicative of sensory disturbance (such as anæsthesia or hyperæsthesia in the paralysed parts; anæsthetic patches in other parts of the body; contrac-

tion of the fields of vision, etc.) are usually present. This point is of great diagnostic importance, as we shall afterwards see when we come to study the hysterical condition. Whereas, in primary sclerosis of the crossed pyramidal tract, sensory disturbances are conspicuous by their absence.

The mode of onset of the paralysis, too, is in some cases distinctive; the onset is always slow and gradual in primary sclerosis of the crossed pyramidal tract; it is sometimes, though by no means always, sudden in hysterical paraplegia, and may directly follow some obvious exciting cause, such as emotional excitement.

But notwithstanding these reservations, a careful and intelligent observer is not likely to mistake primary spastic paraplegia for hysterical paraplegia, except in the early stages of the disease and in those cases in which the patient is a young woman.

In those cases in which hysterical symptoms and sensory disturbances are absent, and in which typical ankle-clonus is well marked, the more serious affection should always be suspected, even although the rigidity and tension are not marked. In doubtful cases, the more serious affection should always be suspected and a guarded prognosis given. It is not of course a satisfactory thing to give a hesitating opinion. None of us like to admit that a case beats us. Some people never make this admission; they make a point of giving a positive opinion as to the nature of every case which they come across; but they are the people who know least about medicine, and who make the biggest mistakes. The most accurate, careful and experienced clinicians often find it necessary to hesitate and give a guarded opinion; to go back again and again to the case, to examine it and re-examine it, and to think over it before coming to a definite conclusion as to its nature. In cases which seem difficult and complicated at the first visit, it is always a good plan to let a night pass and to examine the case again before giving a definite opinion. Even if one does not actively and consciously think about the case, the facts go on simmering, as it were, in one's mind. At the second visit, new facts are in some cases elicited, and the points of the case are often grasped and appreciated in a way which seemed impossible at the first examination. In many cases, the difficulties seem to have disappeared when one sees the case a second time; and it is not always easy

or possible to explain to oneself the mental process by which this satisfactory result is arrived at. I repeat, in doubtful cases, give a guarded opinion; examine the patient again and again, until you make up your mind; and as long as you are in a condition of uncertainty, watch and wait. It is seldom of course necessary or advisable to tell all this to the patient; and you must treat the patient while you are forming an opinion. I do not require to tell you that it would be a very grave mistake to say to a young woman who was going in for primary spastic paraplegia, 'There is nothing the matter with you except hysteria'; but I may take this opportunity of telling you that even if the case is one of hysterical paralysis, you should on no account make such a statement. Patients do not like to be told, 'There is nothing the matter with you but hysteria.' If you put the matter in that way, you will not be likely to cure the paralysis, and you will perhaps lose the patient, and possibly the whole connection. And here let me say that if you wish to make a big practice, the connection is more important than the individual patient; but in order to get the connection you must please and cure your individual patients.

The differential diagnosis of primary sclerosis of the crossed pyramidal tracts and of disseminated (cerebro-spinal) sclerosis.—In typical cases of cerebro-spinal sclerosis, there is no difficulty, but in the early stages of those cases of disseminated sclerosis in which the lesion is limited to the spinal cord it may be very difficult or impossible to distinguish the two conditions. Thirteen years ago I saw with Dr. Sinclair of Dundee a patient who presented all the characteristic symptoms of primary spastic paraplegia, and who presented none of the characteristic symptoms of cerebro-spinal sclerosis. A few months ago the patient again consulted me; all the typical symptoms of disseminated cerebro-spinal sclerosis were then present. I have seen several other cases of the same kind, and I have been so much impressed with them that I always think of disseminated cerebro-spinal sclerosis when I meet with what appears to be a case of primary spastic paraplegia. When I come to describe disseminated cerebro-spinal sclerosis I will refer to this diagnostic problem in more detail.

The differential diagnosis of primary sclerosis of the crossed pyramidal tracts and of transverse myelitis above the lumbar enlargement, will be more appropriately considered after I have described

myelitis. I may, however, say that the slow and gradual mode of onset, the absence of sensory disturbances (anæsthesia, girdle sensation, etc.) of marked paralysis of the bladder or rectum, and of bedsores, are in favour of primary sclerosis of the crossed pyramidal tracts.

The differential diagnosis of primary sclerosis of one crossed pyramidal tract and of cerebral hemiplegia.—Primary sclerosis of the crossed pyramidal tract limited, or chiefly limited to one side of the cord might easily be mistaken for hemiplegia of cerebral origin; the difficulty is theoretical rather than practical, for such limitation is exceedingly rare. Nor is the distinction really difficult. The onset of cerebral hemiplegia is usually rapid; in those cases in which the paralysis is slowly developed, characteristic symptoms of a 'coarse' cerebral lesion, such as tumour, are generally present. Again, in cerebral hemiplegia the arm is usually more affected than the leg, and the face is usually involved; while in primary sclerosis of the crossed pyramidal tract the face is not involved, and the leg usually suffers more than the arm.

The differential diagnosis of combined spinal lesions.—Hitherto I have been speaking of those cases in which the sclerosis is limited to the crossed pyramidal tracts; but, as I have more than once pointed out, it is by no means uncommon to find other tracts or areas implicated. I shall return to the differential diagnosis of these combined lesions when I come to describe progressive muscular atrophy, amyotrophic lateral sclerosis, and postero-lateral sclerosis.

Prognosis.—Let us now turn to the prognosis. The course of primary sclerosis of the crossed pyramidal tracts is, as we have seen, usually very chronic; cases are occasionally met with in which the disease has lasted for twenty, twenty-five, or even thirty years. The lesion does not of itself tend to shorten life; it does not predispose to the occurrence of any visceral complication; and grave complications of a urinary kind, such as septic cystitis, pyelitis and surgical kidney are, as I have previously stated, comparatively speaking very rarely developed. The prognosis as regards the duration is therefore in most cases favourable. Let me say in passing that chronic lesions of the spinal cord usually kill either—(a) by the extension of the lesion upwards

to the medulla oblongata and the consequent involvement of the vital (respiratory and cardiac) centres; (*b*) by producing paralysis of the respiratory muscles—even a slight degree of respiratory paralysis is serious, for intercurrent attacks of bronchitis or pneumonia are, under such circumstances, very likely to prove fatal; or (*c*) by the development of grave bladder and urinary complications, the result of paralysis of the bladder and septic cystitis. (*d*) Death is in many cases also due to phthisis or other intercurrent complications. I am speaking, you will observe, of the chronic affections of the spinal cord; acute lesions may kill in other ways.

Further, you must remember that in this disease (primary sclerosis of the crossed pyramidal tracts), there is no pain and little or no suffering. Now in locomotor ataxia, which is also due to a chronic sclerotic lesion, the pain and suffering are often very great. In locomotor ataxia, the frequent occurrence of severe attacks of pain may seriously impair the general health, and, what is perhaps more important, may cause profound mental depression which exerts a very prejudicial effect upon the course of the disease, or may lead to the establishment of the morphia habit.

The prognosis as regards cure is unfavourable. In my own experience, few cases of primary sclerosis of the crossed pyramidal tract are cured, once the lesion is definitely and distinctly established. It is only, however, right to say that some authorities take a more hopeful view, and state that the disease is not unfrequently cured. It is true that, in some cases, improvement occurs from time to time, and that, in others, the progress of the lesion is arrested; but arrest is not cure. The earlier the case comes under observation, the better is the chance of arrest and (?) cure.

In considering the prognosis of spastic paraplegia it is important to draw a distinction between those cases in which the lesion of the crossed pyramidal tract is primary and those in which it is secondary; for in some forms of secondary spastic paraplegia the prognosis is more hopeful than in the primary variety. In cases of spastic paraplegia due to pressure on the cord, the paralysis may completely pass away provided that the cause of the pressure can be removed and cured. In many cases of syphilitic myelitis, too, the paraplegia may entirely or

largely disappear under appropriate treatment. But I need not go into details. We shall consider the prognosis of the different forms of myelitis and pressure paraplegia in subsequent lectures.

When a patient comes before you with primary sclerosis of the crossed pyramidal tract, I would not advise you to hold out much hope of recovery; but you may confidently assure him that the disease is not dangerous to life, and that at the worst (if the lesion continues to progress) the downward course will in all probability be slow. Further, if the disease is in an early stage, you may perhaps hold out some hope that under appropriate treatment it may possibly be arrested. This, notwithstanding the opinion of some authorities, is as far as I myself would feel disposed to go in encouraging the patient's hopes.

Treatment.—Since the hope of completely curing the disease is very small, the main objects of treatment are to arrest the development of the sclerotic process, and to guard the patient against all conditions which are likely to be injurious and to hasten the progress of the disease. The general health must be kept in the best possible state of efficiency. The patient should be carefully protected from cold, because external cold not only makes the stiffness and rigidity worse, but probably has a prejudicial effect upon the cord lesion. You will find that almost all patients who are suffering from chronic cord disease bear cold badly. When the patient can afford it, he should pass the winter and cold spring months in a warm genial climate, such as Egypt, the Riviera, the Canaries, etc. Over-fatigue and over-exertion should also be avoided. Some authorities believe that excessive muscular work is in some cases the cause of the disease; and there is, I think, good reason to suppose that muscular exercise which causes a marked increase in the rigidity, which produces distinct fatigue and aching pains in the muscles is apt to be injurious; but a certain amount of exercise is usually beneficial. You must remember that in spastic paraplegia muscular exercise is not required for the purpose of keeping up the nutrition of his muscles. In spastic paraplegia, the muscles are being constantly thrown into reflex contraction—constantly exercised—irrespective of the efforts of the will. This reflex exercise is often more than is required; and here let me say that one important point in the treatment of primary spastic paraplegia is to avoid anything

which unduly irritates and excites the abnormally irritable reflex centres of the cord. In speaking of the treatment of poliomyelitis anterior acuta, I told you that some people when they meet with a case of paralysis at once prescribe strychnine and electricity as a matter of routine. Now in this disease (primary spastic paraplegia) strychnine and the faradic current ought not to be prescribed. In most cases of spastic paraplegia they do harm. I am speaking of those cases in which there is no muscular atrophy. The faradic current is applied to paralysed muscles with the object of exciting muscular contraction and of maintaining or restoring their nutrition. Now in primary spastic paraplegia the muscular nutrition is good. The faradic current is consequently not required. It is apt to do harm, for it increases the spasm and by stimulating the sensory nerves in the skin and muscles produces reflex irritation of the already over-irritable nerve cells in the anterior horn of the spinal cord.

Strychnine still further excites the over-irritable nerve cells and increases the spasm. It is a good general rule to remember that strychnine and the faradic current are chiefly useful in those forms of paralysis in which there is flaccidity and atrophy; whereas they are uncalled for and often injurious in those cases of paralysis in which the nutrition of the muscles is well preserved and in which there is rigidity and tension and marked exaggeration of the deep reflexes. But there are not a few cases in which spasm and atrophy are combined. Now, in these cases, small doses of strychnine or nux vomica with hydrobromic acid are often, I believe, beneficial. I may say in passing that in some cases of functional paralysis, the nerve cells are in a condition of irritable weakness—cases in which the knee-jerks are exaggerated and in which the muscles are feeble and flaccid—strychnine is a most valuable remedy; but in such cases there is no persisting rigidity and tension.

In cases of primary spastic paraplegia, the galvanic current, applied to the spinal column (i.e. through the spinal cord), is thought by some authorities to be beneficial. Others are doubtful of its utility. But I need not discuss this question in detail. We have already considered it in our study of poliomyelitis anterior acuta (see page 100). I then told you that, provided that the constant current is used cautiously and

judiciously, it is not likely when applied to the spinal column to do harm, and it may possibly do good.

After what I have just said you may perhaps be disposed to ask whether there are any local or drug remedies which are useful in this disease. To this question I would reply that local warmth (warm baths) and gentle massage seem in some cases to be beneficial; that counter-irritation (blistering the back, the application of the actual cautery to the spine) and dry cupping over the back seem in some cases to be useful; and that internally, arsenic, nitrate of silver, hydrobromic acid and bromide of potassium are useful drugs. Ergot and belladonna have also been recommended. In those cases in which the patient has had syphilis (but this is, as we have seen, rare), iodide of potassium or mercury should of course be tried; but even when there is a definite history of previous syphilis, these remedies seldom prove efficacious, for the previous syphilis is often a mere coincidence, not a distinct cause of the disease.

Arsenic and nitrate of silver are probably the best remedies which we possess, and I am confident that I have seen improvement in some cases of sclerosis in which they were systematically administered. Hydrobromic acid and bromide of potassium do not, so far as I know, exert any beneficial effect upon the lesion itself, but they certainly in some cases allay irritation and spasm. I prefer hydrobromic acid to bromide of potassium since it is less depressing.

The actual cautery is a severe remedy, and must of course be applied with the aid of an anæsthetic; for if it is to do any good, the skin must be freely burned along an extensive tract on each side of the spinal column. The actual cautery is, I think, chiefly useful in those cases of cord disease in which there is meningitis. I have seen it do good in some cases in which spastic paraplegia was the result of myelitis. Spastic paraplegia due to primary sclerosis of the crossed pyramidal tracts is so incurable that one is glad to employ any means which seem likely to hold out even a chance of improvement. If I were so unfortunate as to be affected with the disease I would assuredly have the actual cautery applied.

Dry cupping over the spine is, I think, chiefly useful in the earlier stages of the case, but is far less efficacious in primary spastic paraplegia than in myelitis.

In the later stages of the disease, when the patient is confined to bed, it is important to see that he is kept dry and clean, and that the back and hips are not subjected to continuous and injurious pressure. Although the risk of bedsores is not nearly so great in primary spastic paraplegia as in some other cord affections—more especially myelitis—the back requires attention. In the later stages of every form of paralysis (i.e. when the patient becomes bedridden), it is advisable to place the patient on a water bed, provided of course that it can be procured. This is an important proviso, for water beds are expensive.

Further, in the later stages of the disease the condition of the bladder may require special attention. I shall return to this point in connection with the treatment of myelitis.

LECTURE X

PROGRESSIVE MUSCULAR ATROPHY

TO-DAY, Gentlemen, I propose to direct your attention to progressive muscular atrophy. The synonyms *Wasting Palsy*, *Poliomyelitis Anterior Chronica* (chronic inflammation of the anterior horn of the grey matter), *The Aran-Duchenne Type of Progressive Muscular Atrophy*, and *The Spinal or Myelopathic Form of Progressive Muscular Atrophy* have been given to this disease.

The Aran-Duchenne type (the spinal form) of progressive muscular atrophy is in most cases a very chronic disease. It essentially depends upon a slow and gradual destruction of the multipolar nerve cells in localised segments of the spinal cord. And this leads me to say that in other cases of diffuse and progressive atrophy there is no discoverable cord lesion. The terms *idiopathic* muscular atrophy and *myopathic* muscular atrophy have been applied to these cases. I shall return to this subject later. Let me first describe the common, spinal, form of the disease.

Pathological Anatomy.—I have said that the essential pathological substratum of the ordinary form of progressive muscular atrophy is a slow and gradual destruction of the multipolar nerve cells in the anterior horn of the spinal cord. In some cases, the lesion is *limited* to the region of the anterior horn, at least so far as the clinical symptoms enable us to judge. It is for this reason that the disease is included under the system lesions or diseases of the spinal cord. In other cases, in which the symptoms are characteristic of progressive muscular atrophy, the crossed pyramidal tracts (sometimes also the direct pyramidal tracts) are also implicated. The indications of this sclerosis are not always very apparent during life; but it is said that in many

of the cases in which there are no symptoms indicative of a lesion of the crossed pyramidal tracts during life, the pyramidal tracts are distinctly sclerosed after death. Indeed some authorities now suppose that the sclerosis of the crossed pyramidal tracts is not merely an associated lesion or complication, but that it is an essential and constant feature of the disease. The question is still *sub judice* and further observations are perhaps required before a definite opinion can be pronounced regarding it; but, so far as my own observation enables me to judge, cases are undoubtedly met with in which there is absolutely no sclerosis discoverable on microscopic examination in the crossed pyramidal tracts. For our present (practical) purpose (and perhaps in the present position of our knowledge) it will probably be best to look at the question from the clinical rather than from the pathological side.

If then we look at the matter from the standpoint of the clinician, it must, I think, be admitted, *firstly*, that in some cases of progressive muscular atrophy there is no definite clinical evidence (in the form of spastic symptoms and exaggeration of the deep reflexes either in the upper or lower extremities) of any sclerosis in the crossed pyramidal tracts during life; and *secondly*, that in other cases in which the characteristic symptoms of progressive muscular atrophy are well marked, very definite indications of a lesion in the crossed pyramidal tracts (great exaggeration of the knee-jerks, ankle-clonus and perhaps rigidity and tension of the muscles either in the upper or the lower extremities) are present.

The illustrious Charcot attached great importance to these clinical differences. He applied the term *amyotrophic lateral sclerosis* to some of the cases of progressive muscular atrophy in which there is a well marked lesion of the crossed pyramidal tracts. He supposed that in amyotrophic lateral sclerosis the lesion commences in the crossed pyramidal tracts and, after a time, extends on to the anterior horn. According to Charcot, the lesion of the anterior horn in amyotrophic lateral sclerosis is secondary, or, as he termed it, *deuteropathic*; whereas in progressive muscular atrophy, the lesion of the anterior horn is primary or *protopathic*.

But of late years the correctness of this view has been called in question. As I have already pointed out, some leading

authorities now believe that in these combined cases the lesion in the crossed pyramidal tract is coincident with and due to the same cause as the lesion in the anterior horn. According to this view, there is no real distinction (no absolute pathological line of demarcation) between progressive muscular atrophy and amyotrophic lateral sclerosis; they are merely varieties of the same disease; the typical forms which seem to be distinct run insensibly one into the other and are connected by intermediate and less typically differentiated varieties.

But whether we accept this view or not—and personally I do not as yet see my way to accept it—it is, I think, advisable for clinical purposes to differentiate the two varieties and to divide cases of progressive muscular atrophy into two great groups, viz., (1) Cases in which the lesion in the anterior horn is the only or the predominant lesion and the muscular atrophy is the only or the predominant symptom; and (2) Cases in which definite clinical indications of a lesion in the crossed pyramidal tract (muscular rigidity and tension or marked exaggeration of the deep reflexes) are also present. The first group comprises the ordinary (spinal) form of progressive muscular atrophy; the second group the condition which Charcot termed amyotrophic lateral sclerosis. As I have already stated, the two varieties run indefinitely one into the other; and it is not always easy or possible to say whether a given case should be placed in the first or the second group. It is quite common, for example, in the ordinary typical form of progressive muscular atrophy to find the knee-jerks markedly exaggerated and ankle-clonus present, but no definite rigidity or tension in the muscles either of the upper or the lower limbs.

The character of the lesion.—The points which I wish chiefly to emphasise are:—that the spinal form of progressive muscular atrophy is essentially due to a slow and gradual destruction of the multipolar nerve cells in the anterior cornua of the spinal cord; that in many cases, the crossed pyramidal tracts are also sclerosed; and that in some cases, the sclerosis of the crossed pyramidal tracts is so marked that the symptoms which it produces form a conspicuous feature of the clinical picture.

The question has been much debated whether the lesion of the nerve cells is degenerative or inflammatory in character. In some cases, subacute or chronic inflammatory changes in the

grey matter of the anterior horn are undoubtedly present. The presence of such inflammatory changes in the anterior horn does not necessarily prove that the primary lesion of the nerve cells is inflammatory; for inflammatory changes are frequently developed in tissues which are degenerated and diseased. In the present state of our knowledge it is perhaps impossible to pronounce a definite and dogmatic opinion and to say whether the Aran-Duchenne form of progressive muscular atrophy is the result, on the one hand, of a degenerative atrophy of the multipolar nerve cells with secondary inflammatory changes in the neuroglia, or, on the other, of a chronic parenchymatous inflammation of the anterior horn of grey matter, the essential feature of which is a destruction and degeneration of the multipolar (motor) nerve cells.

The mode of extension of the lesion.—In the next place, I must ask you to note that the lesion is not diffused all through the spinal cord; it is limited to certain definite regions and segments. This is a most important clinical characteristic of the spinal (Aran-Duchenne) type of progressive muscular atrophy.

In the great majority of cases, the lesion commences in the lower cervical or first dorsal segments. The nerve cells connected with the small muscles of one or both hands are usually first involved. In this respect, progressive muscular atrophy contrasts remarkably with poliomyelitis anterior acuta. In that disease, the lumbar is much more frequently affected than the cervical enlargement and the paralysis consequently affects the muscles of the legs much more frequently than those of the arms.

In many cases the atrophy commences in the thenar or hypothenar muscles of the right hand. The movements of the fingers and thumb are more highly specialised than any other of the limb movements; and you will remember that mechanisms which are very complicated in structure and highly specialised in function are easily deranged. It would appear, too, that degenerative processes are more apt to develop in the neuro-motor mechanisms which are complex and highly specialised than in the neuro-motor mechanisms which are simple in structure and more automatic in function.

In exceptional cases the atrophy seems to commence in the muscles of the back or lower extremities. But this mode of development perhaps requires further verification. It is only of

recent years that a definite distinction has been drawn between the spinal and idiopathic forms of progressive muscular atrophy; and it seems certain that in the great majority of cases of the Aran-Duchenne type, the small muscles of the hand are first affected. I shall return to this point in connection with the clinical history.

In the great majority of cases, the lesion at its commencement seems chiefly to involve the nerve cells in one, usually the right, anterior horn; consequently the atrophy is, at its commencement, usually more marked, or only marked in the small muscles of one, usually the right, hand.

It usually next involves the nerve cells in the opposite half of the segment which was first affected, i.e. when the small muscles of one (say the right) hand are first involved, the small muscles of the opposite (the left) hand are next affected. But this is not always the case. In some cases, other half segments on the same side of the cord (cervical region) are next affected; in other words, in some cases the small muscles of one (say the right) hand are first involved, and the muscles of the forearm or deltoid on the same (the right) side—not the muscles of the opposite (the left) hand, as is usually the case—are next implicated.

The next point in the pathology which I would ask you to note is that all of the nerve cells in the particular half of the segment which is first affected are not simultaneously involved. The lesion picks out, as it were, some nerve cells, and after slowly and gradually destroying them, passes on to, picks out and destroys other nerve cells. The destructive process is slow. The individual nerve cells which are first affected are slowly destroyed. Months, it may be years, elapse before all the nerve cells in the area of the cord which is first involved are all destroyed; in fact, the destruction is rarely if ever complete; some of the nerve cells usually remain.

The results of this slow and gradual destruction is the production of muscular weakness, atrophy, and ultimately of paralysis of the muscles connected with the affected segments of the cord. The weakness and muscular atrophy are, you will observe, slowly and gradually developed. In this respect progressive muscular atrophy contrasts remarkably with poliomyelitis anterior acuta. In that disease the paralysis is rapidly developed, the maximum

extent of the paralysis is at once produced, and, in the great majority of cases, some of the initial paralysis disappears after the acute inflammatory changes have subsided. It is the exact reverse in progressive muscular atrophy. In that disease, the lesion commences in a very insidious manner, slowly and gradually destroys a few nerve cells, produces little or no obvious derangement at first, but gradually extends and finally involves almost all of the nerve cells in the affected segment, and it may be ultimately produces complete atrophy and loss of power.

Further, in poliomyelitis anterior acuta the lumbar is more frequently affected than the cervical enlargement; whereas in progressive muscular atrophy the cervical enlargement is almost invariably affected, and the lumbar enlargement usually escapes. Again, poliomyelitis anterior acuta is essentially a disease of the child, whereas progressive muscular atrophy is essentially a disease of the adult. A further point of distinction is this, that in poliomyelitis anterior acuta the lesion remains limited to the segments of the spinal cord which were first attacked; but in progressive muscular atrophy the lesion tends to spread to other segments until it may ultimately involve almost every segment in the cervical enlargement. Indeed, it not unfrequently happens that the dorsal and sometimes the lumbar segments are ultimately invaded; in many cases the multipolar (motor) nerve cells in the medulla oblongata are finally implicated.

From these statements you will see that the two diseases present a remarkable contrast in respect to the pathological character of the lesion and the clinical features and course.

Naked eye appearances of the spinal cord.—On examining the cord from a case of progressive muscular atrophy, the most obvious alteration is the grey and wasted condition of the anterior nerve roots. In one typical case in which I obtained a post mortem a few years ago, the contrast between the atrophied anterior and the normal posterior nerve roots was most striking, not only in the cervical, but in the lumbar region of the cord. In that case the lower extremities were markedly affected. Transverse sections of the cauda equina, after hardening, showed this contrast in a very remarkable way.

With the exception of the grey and atrophied condition of the anterior nerve roots there is little or nothing to be seen with the naked eye. In some cases the grey matter of the anterior

horn is said to be softer than normal. But here let me say that softening is often found (or said to be present) after death, in cases in which the spinal cord was perfectly healthy during life. In some of these cases, the softening is a post mortem (decomposition) change; in others it is due to injury (crushing and squeezing) inflicted by a maladroit pathologist. The more inexperienced the pathologist, the more frequent the softening. It is not always an easy matter to say whether the consistency of the cord is normal or not. I do not myself attach much importance to the mere naked eye characters of the cord (its consistency, the presence of softening, etc.), unless indeed the post mortem is made by an experienced pathologist.

Microscopical appearances of the spinal cord.—On examining properly hardened sections the essential change is seen to be an atrophy and in the advanced stages of the disease it may be the disappearance of almost all of the multipolar nerve cells in the anterior horn of the segment or segments of the cord which are chiefly implicated (see Fig. 42). The atrophy seems in some cases to be a simple atrophy; the nerve cells are shrivelled, smaller and less plump than normal; they stain badly; their processes are fewer in number than normal, and in the advanced stages of the disease they may have entirely disappeared. In some cases, it is said that the nuclei still remain in the centre of the atrophied cells. In many cases an excess of pigment is deposited in the shrivelled cells. In short, the essential change in the nerve cells is atrophy, pigmentation, and degeneration.

The minute nerve fibres and the axis-cylinder processes of course share in this degeneration.

The connective tissue (neuroglia) in which the nerve cells are embedded is usually increased in amount (sclerosed), and the connective tissue cells (the Deiters' cells) enlarged. The blood vessels in the affected area are often dilated, their walls thickened, and their hyaline sheaths larger and more capacious than normal. I may take this opportunity of saying that it is a very difficult thing to give a decided opinion as to the exact condition of the connective tissue of the grey matter.

In cases which run an unusually rapid course, fatty granules may be present in the nerve cells or in the spaces which were formerly occupied by nerve cells. But it is doubtful if this fatty change occurs in the ordinary typical (chronic) cases.

In those parts of the cord in which the lesion is less advanced, many healthy nerve cells may still be seen.

The anterior nerve roots connected with the less severely affected parts of the cord do not present the extreme grey

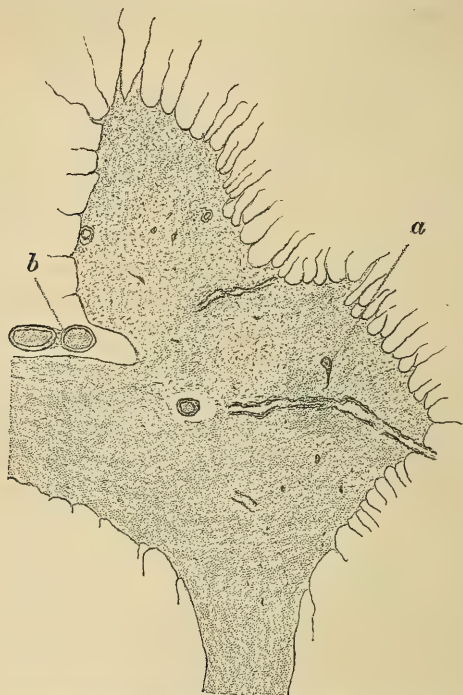


FIG. 42.—*Transverse section through the anterior horn of the spinal cord in a case of progressive muscular atrophy.*

The whole anterior horn of grey matter is shrunken.

The nerve cells have almost entirely disappeared; one cell of normal size and one or two atrophied cells only remain. The blood vessels are much dilated. Under a high power the grey matter of the anterior horn is found to be sclerosed; numerous small corpuscles (leucocytes), which are probably indicative of the inflammatory character of the lesion, are scattered through the grey matter and especially around the dilated vessels.

The letter *a* points to the only large nerve cell in the section. The letter *b* points to dilated blood vessels.

atrophy which the nerve roots connected with the lower part of the cervical enlargements usually exhibit.

The degeneration extends through the nerve roots and peripheral motor nerves down to the muscles.

The naked eye and microscopical appearance of the muscles.
—In advanced cases the muscles are pale and fawn-coloured;

they may look as if they were almost entirely composed of fat. The muscular wasting seems to be essentially a simple atrophy. So far as is known, enlargement of the muscular fibres which is such a conspicuous and characteristic feature of many cases of the myopathic (idiopathic) forms of muscular atrophy does not occur.

In advanced stages of the disease, microscopical examination

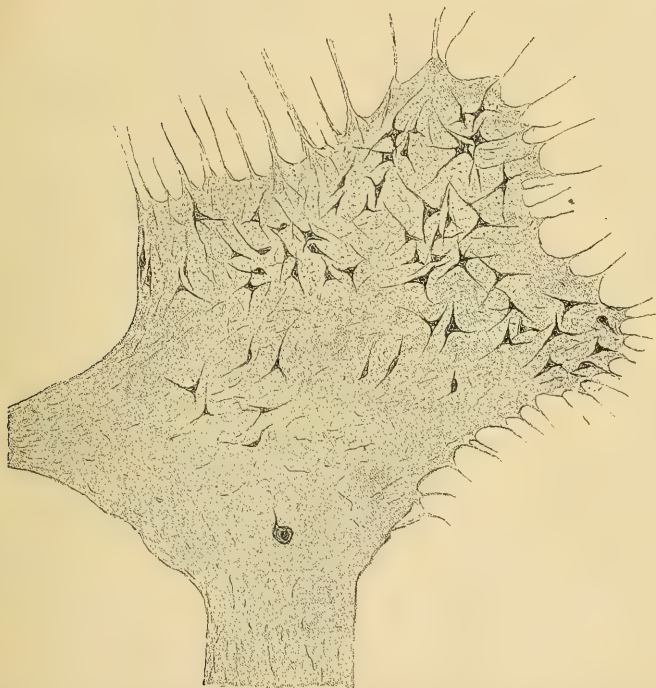


FIG. 43.—*Transverse section through the anterior horn of a healthy spinal cord, for comparison with Fig. 42.*

shows that the muscular fibres have almost entirely disappeared. In the earlier stages, some of the muscular fibres are found to be healthy; others present various stages of atrophy. As I have already told you the essential change seems to be a simple atrophy; muscular fibres which are markedly atrophied and narrowed may still retain their transverse striation. In the advanced stages, the transverse striation may completely disappear, and the muscular fibres may be infiltrated with granular

and fatty globules. The muscle nuclei may be increased in number. Some authorities state that some of the muscular fibres are sometimes affected with a colloid or hyaline degeneration. The spaces between the individual muscular fibres may be occupied by an excess of connective tissue, dilated blood vessels and a few fat cells; but the increase of the connective tissue is not, as a rule, so marked as it is in many cases of poliomyelitis anterior acuta, and is always far less apparent than in pseudohypertrophic paralysis.

As I have already more than once told you, the crossed pyramidal tracts (and, I may add, the anterior pyramidal tracts), are in many cases sclerosed. Indeed, there may be some increase of the connective tissue through the whole transverse thickness of the cord. Nevertheless, the posterior horn of grey matter, the posterior roots and posterior columns are, practically speaking, always healthy.

In many cases the sclerosis of the crossed pyramidal tract is only slight; and in some cases it is entirely absent.

In cases of amyotrophic lateral sclerosis, the sclerosis of the pyramidal tracts extends throughout the whole length of the cord, and, indeed, higher up, into the medulla, pons Varolii, and it may be even through the crus cerebri and internal capsule to the cortex itself.

In those cases in which there is a lesion both in the anterior cornua and the crossed pyramidal tracts the sclerotic lesion in the crossed pyramidal tracts is clearly not the cause of the lesion in the anterior horn; for, as I have already more than once pointed out, cases are frequently met with in which, so far as the clinical evidence shows, the lesion seems to be entirely limited to the anterior horn and in which there are no symptoms indicative of a lesion in the lateral columns. Further, we know that in primary spastic paraplegia and in secondary degeneration of the crossed pyramidal tracts, the lesion in the crossed pyramidal tracts is not, unless in quite exceptional instances, attended with any degeneration of the multipolar nerve cells in the anterior horn of grey matter.

M. Marie has advanced the view that the lesion in the crossed pyramidal tract is secondary to the lesion in the anterior horn. The fact that in poliomyelitis anterior acuta, acute destruction of the nerve cells of the anterior horn and acute inflammation of

the anterior horn is not followed by sclerosis in the crossed pyramidal tracts, seems strongly opposed to this theory. The arguments which M. Marie advances in support of his theory are:—That the degeneration of the pyramidal fibres does not extend to the pons Varolii and medulla oblongata; that the sclerosis in the lateral column is not strictly limited to the area of the crossed pyramidal tract; and that the direct pyramidal tract is not affected.

A third view supposes that the two lesions are not directly related as cause and effect, but that they are simultaneously developed, and are probably due to a common cause acting upon the whole motor tract. If this is so, we would not expect to find any direct relationship, as regards severity, between the muscular atrophy (the clinical manifestation of the lesion in the anterior horn) and the rigidity and tension (the clinical manifestations of the lesion in the pyramidal tracts); and such is the case.

I have already stated that in some cases of progressive muscular atrophy, the lesion extends to, and involves, the motor nerve cells of the medulla oblongata, and it may be of the pons Varolii. This is one of the most important points connected with the morbid anatomy and the clinical history of the disease; for involvement of the cardiac and respiratory centres in the medulla oblongata is a frequent cause of death. In some cases in which progressive muscular atrophy in the limbs and trunk and bulbar symptoms are combined, the spinal nuclei are first involved; in others, the bulbar nuclei are first affected; while in a third group of cases the spinal and bulbar nuclei are simultaneously affected. I repeat, that so far as we know, the lesion in the medulla oblongata, which is the pathological substratum of glosso-labio-laryngeal paralysis is identical with the lesion in the cord which is the pathological substratum of progressive muscular atrophy. In both cases the lesion is a degeneration of the motor (multipolar) nerve cells. In both affections the pyramidal tracts are frequently involved.

It was at one time supposed that progressive muscular atrophy was due to a lesion of the sympathetic; but we now know that this view was a mistaken one.

Further, the theory which was strenuously supported by Friedreich, viz., that the primary cause of the disease is situated in the muscles, has been proved to be erroneous. As I have

already sufficiently pointed out, pathological investigation has conclusively proved that the Aran-Duchenne type of progressive muscular atrophy is due to a lesion of the multipolar nerve cells of the anterior cornua of the spinal cord.

Etiology.—Progressive muscular atrophy usually commences between the ages of thirty and fifty. The disease is very rare before the age of twenty-five, and exceedingly rare in childhood. I refer to the typical spinal (Aran-Duchenne) form of the disease. The idiopathic (myopathic) forms of muscular atrophy which in the great majority of instances commence in childhood or youth, must, as I have already pointed out, be carefully distinguished from the common spinal form of progressive muscular atrophy which we are now considering.

Progressive muscular atrophy comparatively seldom commences after the age of fifty. In this respect, it presents a remarkable difference from glosso-labio-laryngeal paralysis, for that disease rarely develops before the age of fifty.

Progressive muscular atrophy is much more common in men than in women. It seems to attack the lower orders of society more frequently than the upper. At one time it was supposed that the disease is often directly inherited, but we now know that this view was a mistaken one. The Aran-Duchenne type of progressive muscular atrophy is very rarely, some authorities say never, directly inherited. In this respect the spinal or myelopathic form of progressive muscular atrophy differs very markedly from the idiopathic or myopathic forms of muscular atrophy (pseudo-hypertrophic paralysis, the juvenile form of Erb and the allied facio-humeral type of Landouzy and Déjerine), in all of which and in the peroneal type of progressive muscular atrophy, which is probably myelopathic rather than myopathic in nature, the disease can often be traced through several succeeding generations.

We know little as to the exact cause of the degenerative or inflammatory lesion which is the pathological substratum of the ordinary (spinal) form of progressive muscular atrophy. Many conditions have been blamed in individual cases, but great caution is necessary in drawing conclusions on this point. The onset is so insidious that in many cases the disease has already been in existence for some time before the muscular atrophy is

noticed, and before the conditions which are blamed as the cause come into effect.

There seems good reason to suppose that excessive muscular effort is sometimes the cause, or at least an exciting cause, of the disease. You can easily understand that if the nerve cells have a tendency to degenerate, prolonged and excessive muscular effort, which throws a strain upon them, will be likely to excite or hasten the development of the degeneration. I have seen several cases which seemed to support this view. In one case, for example, the patient, a great pianist, was in the habit of playing the piano many hours each day. In another case, the patient was a blacksmith; in his case the over-use of the muscles of the hand and forearm which the effort of constantly grasping and raising a heavy hammer necessitated, was perhaps the exciting cause of the disease.

Traumatic injury to the back is sometimes blamed, and it is impossible to deny that concussion of the spinal cord and resulting injury to the delicate grey matter of the anterior horn (punctiform ecchymoses) may perhaps in some cases be the cause of the disease. But the cases in which progressive muscular atrophy can, with any degree of certainty or even probability, be attributed to concussion of the cord or direct injury to the back are, so far as my information enables me to judge, exceedingly rare. Further, injury to a limb is occasionally followed by progressive muscular atrophy, and it has been supposed that in such cases the cord lesion is the result of an irritation, or perhaps an inflammation, which extends up the nerves from the seat of the injury to the spinal cord.

I need not say that the disease is often said to be the result of exposure to cold; possibly in some cases this cause is really effective. The disease sometimes develops (but this is, comparatively speaking, very rare) after syphilis. A well marked case of this kind came under my notice quite recently; but I doubt whether the syphilitic factor was more than a mere coincidence. It is possible that in many of the cases in which progressive muscular atrophy was thought to be the direct result of syphilis, the lesion which produced the muscular atrophy was situated in the peripheral nerves and not in the spinal cord. The Aran-Duchenne form of progressive muscular atrophy seems to be very rarely developed after an attack of one of the

specific fevers. In this respect it contrasts remarkably with the peroneal type of muscular atrophy, which, as I shall afterwards have to tell you, seems to be more closely related to this, the spinal, form of progressive muscular atrophy than to the idiopathic (myopathic) forms of muscular atrophy with which it is usually classified. Lead impregnation sometimes causes a diffuse muscular wasting which is readily mistaken for progressive muscular atrophy; possibly in rare cases lead poisoning is an actual cause of the disease.

In many cases, the conditions to which I have just referred as supposed causes are probably mere incidental factors, or at most exciting causes or aggravating conditions. In the present position of our knowledge, it is perhaps impossible to make a more precise statement respecting the etiology than this, viz., that the lesion seems to be a degeneration or chronic inflammation of the motor nerve cells of the spinal cord, and that anything which irritates, exhausts, or throws a strain on the nerve cells (which are predisposed to degenerate or which are already in a commencing state of degeneration) may probably act as an exciting cause of the degeneration, or hasten its progress once it is developed.

In connection with the etiology of the disease, I must not omit to mention the interesting fact that Roger (quoted by Grasset and Rauzier¹), by injecting old cultures of the streptococcus of erysipelas into rabbits, produced a localised myelitis, the essential pathological feature of which was degeneration of the multipolar nerve cells of the anterior cornua of the spinal cord. This experimental lesion was attended with clinical symptoms identical with those of progressive muscular atrophy.

¹ *Maladies du Système Nerveux*, page 622.

LECTURE XI

PROGRESSIVE MUSCULAR ATROPHY (*Continued*)

Clinical History.—In typical cases, progressive muscular atrophy is a very chronic disease. The onset is usually insidious and is unattended with fever or constitutional disturbance. In many cases, the atrophy has already made considerable advance before the patient becomes aware of its presence. This statement, of course, applies to non-professional persons, and especially to unobservant people in the lower ranks of society. Skilled observers (medical men) will be likely to detect the disease in its early stages; indeed medical men and medical students not unfrequently imagine (quite erroneously) that they are affected with progressive muscular atrophy. I shall return to this point when I come to speak of the diagnosis.

We have seen that according to some authorities the ordinary (spinal) form of progressive muscular atrophy and amyotrophic lateral sclerosis are mere varieties of the same disease. Bearing this opinion (with which I cannot as yet see my way to agree) in view, and admitting that in some cases of progressive muscular atrophy the crossed pyramidal tracts are sclerosed, let us consider the ordinary typical form of the disease, in which the lesion is, for clinical and practical purposes, confined to the anterior cornua of the spinal cord.

Muscular weakness in the hand (for the intrinsic muscles of the hand are in most cases first affected) is usually the first symptom which attracts attention.

In some (but they probably constitute a small proportion of the whole), the development of the disease is attended with myalgic or muscular pains. These myalgic pains may occur at any stage of the disease; they are more frequent in those cases which run an exceptionally rapid course. A more common symptom is a feeling of fatigue after exertion. True myalgic

pains are much less common in progressive muscular atrophy than in some other affections, such as peripheral neuritis, in which muscular wasting is a prominent symptom.

In consequence of the muscular weakness, the patient finds a difficulty in performing certain movements. Thus, as Roberts graphically puts it:—‘The tailor discovers that he cannot hold his needle; the shoemaker wonders he cannot thrust his awl; the mason finds his hammer, formerly a plaything in his hand, now too heavy for his utmost strength; the gentleman feels an awkwardness in handling his pen, in pulling out his pocket handkerchief, or in putting on his hat. One man discovered his ailment in thrusting on a horse’s collar; another, a sportsman, in bringing the fowling-piece to his shoulder.’

Speaking generally, the loss of muscular power is proportionate to and coincident with the amount of atrophy which is present (see fig. 44). This is a general statement. Of course, the weakness is not absolutely equal to the atrophy, for before the nerve cells are completely destroyed, the functional activity of the muscular fibres with which they are connected is necessarily enfeebled; but speaking generally, the amount of atrophy is roughly proportionate to the degree of muscular weakness.

In the great majority of cases, the muscular weakness and atrophy are first observed in the upper extremity. The muscles of the thumb (adductor longus pollicis and opponens pollicis) are usually first affected; then the muscles of the hypothenar eminence, and the interossei. In some cases, the muscles of the forearm, the deltoid, or some of the other muscles of the shoulder girdle or upper arm are first attacked; but this is exceptional. It is said that in rare cases the atrophy first develops in the muscles of the back or of the lower extremities. In many cases, the atrophy is already well advanced when the patient consults a doctor.

After the atrophy has developed to a certain degree on one side, it usually affects the corresponding muscles on the opposite side. But this is not by any means always the case. In many cases, the muscular weakness and atrophy are for a time limited to one arm or the muscles which attach the arm to the trunk; a very considerable degree of wasting may be produced in the arm, which is first affected, before the muscles of the opposite limb are involved. In a small proportion of cases, the

muscles of the lower extremities (the glutei, the muscles of the thigh, or the muscles below the knee) are first attacked. In some cases, the thoracic or trunk muscles are affected in the early stages of the disease; in rare cases, they are first involved.

The muscles of the thorax and of the neck are in some cases ultimately implicated; the diaphragm may be invaded as well

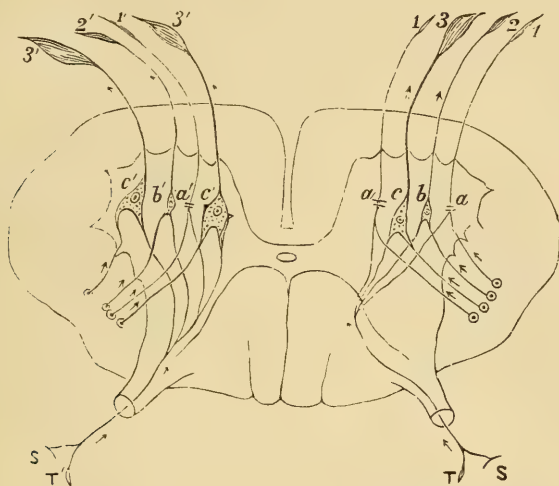


FIG. 44.—Diagrammatic representation of the symptoms which result from slow destruction of the multipolar nerve cells of the anterior cornu.

On the left side the disease is in an early stage. One nerve cell (*a'*) is completely destroyed. Its muscular fibre (*1'*) is completely atrophied. Voluntary motor and reflex motor impulses are 'blocked' at the seat of the lesion (*a'*). One nerve cell (*b'*) and its muscular fibre (*2'*) are very much atrophied, but feeble motor and reflex impulses can still pass through the cell to the muscle. Two nerve cells *c' c'* are healthy. Their muscular fibres are of normal bulk, and can be made to contract either by voluntary or reflex impulses.

On the right side the disease is much more advanced. The muscular area is three-fourths degenerated. There is a total 'block' at *a* and *a'*. This condition represents the late stage of progressive muscular atrophy. The atrophy of the muscular fibres is represented as *simple*.

as the intercostal muscles. In the case which is represented in figs. 55 and 56, the muscles of the back and the intercostals were markedly atrophied.

The clavicular part of the trapezius is very rarely affected, so seldom that Duchenne termed it *ultimum moriens*—the last muscle of the upper extremities and trunk to be attacked.

The atrophy very seldom indeed affects the muscles of

expression or the muscles of the tongue; but, as I have more than once stated, the motor nerve cells in the medulla oblongata are in many cases implicated in the later stages of the case, with the production of bulbar symptoms (muscular weakness and atrophy of the lips, tongue, pharynx, etc.).

The muscular weakness and atrophy gradually extend and involve other muscles, until finally almost all the muscles of the upper extremities and many of the muscles of the trunk may be implicated.

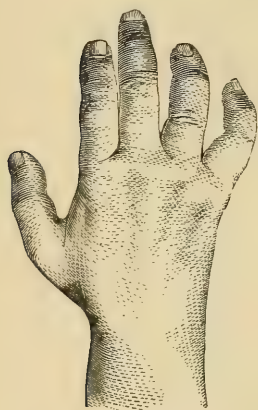


FIG. 45.

The *main en griffe* in progressive muscular atrophy. The interossei and thenar muscles are almost entirely destroyed.—(After Duchenne.)



FIG. 46.

The *main en griffe* which results from paralysis of the interossei in consequence of a traumatic lesion of the ulnar nerve.—(After Duchenne.)

The muscular wasting is the most conspicuous—in fact, in many cases the only—alteration which is apparent. In the hand, the thenar and hypothenar eminences disappear, and depressions, due to the wasting of the interossei, are seen on the back of the hand between the metacarpal bones.

Atrophy of the interossei (the muscles of the forearm being unaffected) causes a peculiar alteration in the position of the fingers, to which the terms ‘clawed hand,’ ‘*main en griffe*,’ have been given (see figs. 45 and 46).

The manner in which this alteration in the position of the fingers is produced is as follows:—

The combined action of the internal and external interossei produces (as Duchenne was the first to demonstrate) a movement of the fingers in which the first phalanx is flexed on the metacarpus, and the second and third phalanges are kept extended; in other words, the action of the interossei is to place the fingers in the writing position. The direction of the tendon of the interossei perfectly explains this contradictory action upon the phalanges; in the first part of its course (from the metacarpal phalangeal articulation to the upper surface of the first phalanx), the tendon is directed obliquely from above downwards and from before backwards (see fig. 47); there is therefore flexion

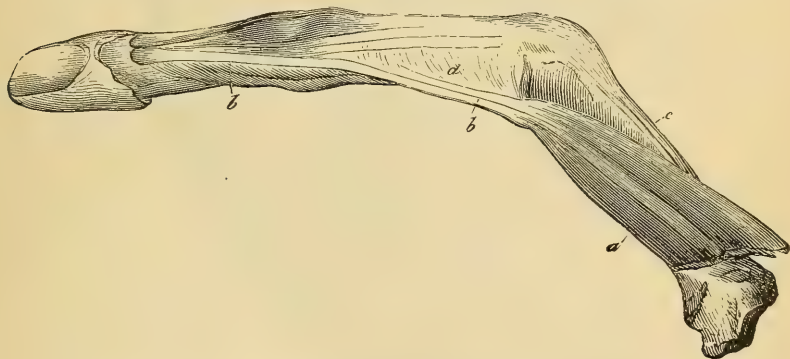


FIG. 47.—Ring finger of the right hand, with its dorsal interosseous or adductor muscle.—(After Duchenne.)

(a) Dorsal interosseous or adductor muscle; (bb) phalangeal tendon of the dorsal interosseous; (c) tendon of the extensor communis; (d) aponeurotic expansion, which unites the phalangeal tendon of the interosseous with the tendon of the extensor.

of the phalanx on its metacarpal bone during contraction of the muscle. The second part of the tendon, which is united by an aponeurotic expansion to the tendon of the common extensor, is placed on the back of the phalangeal articulations; consequently the second and third phalanges are extended on the first phalanx during the contraction of the muscle.

Now, when the interossei are paralysed, the opponent muscles have full play; the result is extension of the first, and flexion of the second and third phalanges; in short, the hand assumes the 'bird-claw' position.

The *main en griffe* or 'bird-claw' hand is not pathognomonic of progressive muscular atrophy; it merely indicates paralysis

of the interossei, a condition which may, of course, be due to a lesion of the ulnar nerve. The appearance of the hand in the two cases (progressive muscular atrophy and paralysis due to a lesion of the ulnar nerve) is, however, somewhat different, a fact



FIG. 48.—*The appearance of the hand (dorsal aspect) after division of the ulnar nerve.*

to which attention was first directed by Duchenne; in progressive muscular atrophy, all the fingers are equally affected; in paralysis of the ulnar nerve, the ring and middle fingers are more particularly involved (see figs. 48 and 49).

The appearance which the forearm presents when its muscles are extensively atrophied is well seen in figs. 50, 51, and 52.

When the deltoid is affected, the roundness and plumpness of the shoulder disappears and the head of the humerus and bony prominences around the joint project conspicuously.

When the trunk muscles are affected in a marked degree,



FIG. 49.—*The appearance of the hand (palmar aspect) after division of the ulnar nerve.*

curvatures of various forms are apt to be produced. Fig. 53 represents a case in which the lumbar muscles were atrophied. In the erect position the back is curved so that a line drawn perpendicularly downwards from the shoulders falls right behind the sacrum; the pelvis is tilted forwards; and there is a compensatory backward curvature of the upper part of the spine,

which entirely disappears when the patient assumes the sitting position; the back is then rounded and projects.

Fig. 54 represents a case in which the abdominal muscles were atrophied. The back is bent backwards by the unopposed lumbar muscles, which were healthy; the abdomen is prominent;



FIG. 50.



FIG. 51.

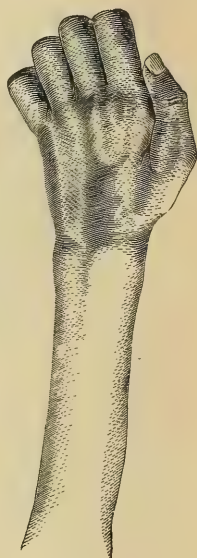


FIG. 52.

FIG. 50.—Hand in a case of progressive muscular atrophy which commenced in the muscles of the thumb.—(*After Duchenne.*)

FIGS. 51 AND 52.—Hands, the muscles of which are almost entirely destroyed, from a case of progressive muscular atrophy, which had become general at the end of two years.—(*After Duchenne.*)

and a line drawn perpendicularly downwards from the shoulders falls well inside the sacrum.

Figs. 55 and 56 represent a typical case of advanced progressive muscular atrophy. The patient was under my observation for many years, and was examined post mortem by Dr. Alexander Bruce and myself. The small muscles of the hand are completely wasted; and the muscles of the forearms and upper

arms have almost entirely disappeared. The only movement which the patient could make with his upper limbs was to swing them about (in a feeble way) like a flail. The muscles of the thorax and back are markedly affected; the muscles

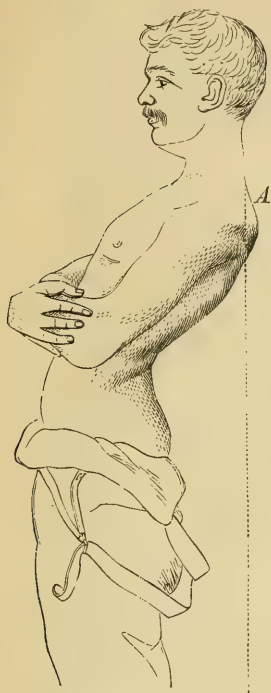


FIG. 53.

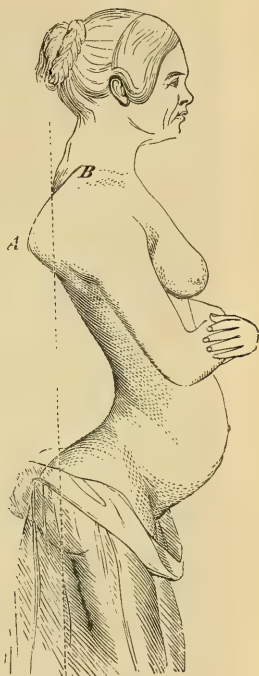


FIG. 54.

FIG. 53.—*Atrophy of the lumbar muscles in progressive muscular atrophy.*—
(After Duchenne.)

In the erect position the back is curved, so that a line, drawn perpendicularly downwards from the shoulders, falls behind the sacrum.

FIG. 54.—*Atrophy of the abdominal muscles in progressive muscular atrophy.*—
(After Duchenne.)

The back is bent backwards by the unopposed action of the lumbar muscles, which are healthy.

of the neck are in some degree involved. The muscles of the lower extremities have almost entirely escaped. I may say in passing that in this very typical case the lesion was confined to the region of the anterior horn; there was absolutely no sclerosis of the crossed pyramidal tracts.

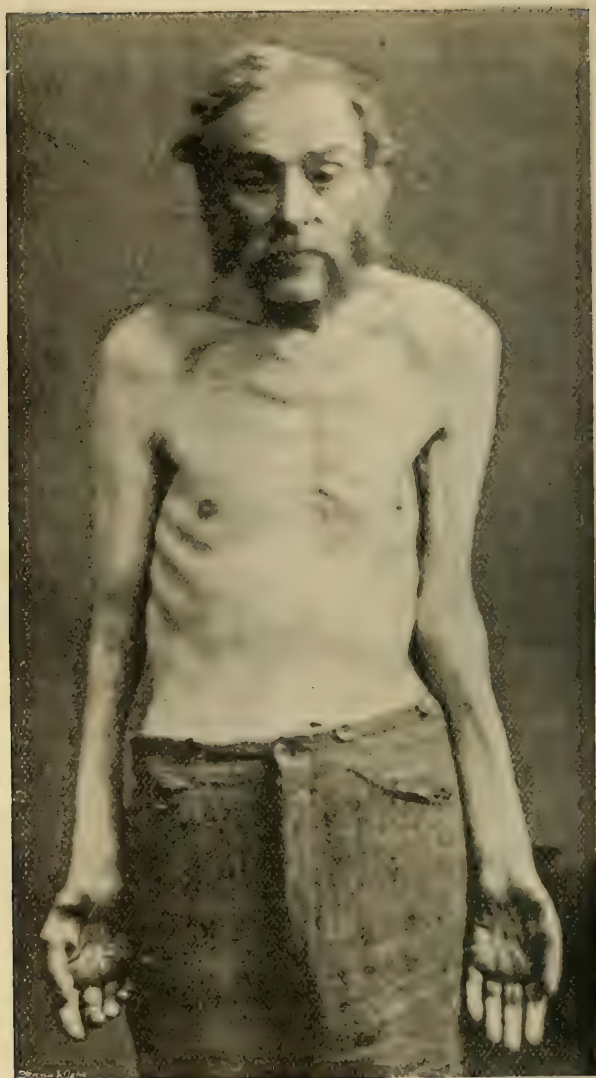


FIG. 55.—*Progressive Muscular Atrophy.*

All of the muscles of the upper extremities are markedly atrophied; the thenar eminences are represented by deep depressions; the muscles of the forearms have completely disappeared; the pectoral muscles are markedly affected. The deltoids are well preserved.

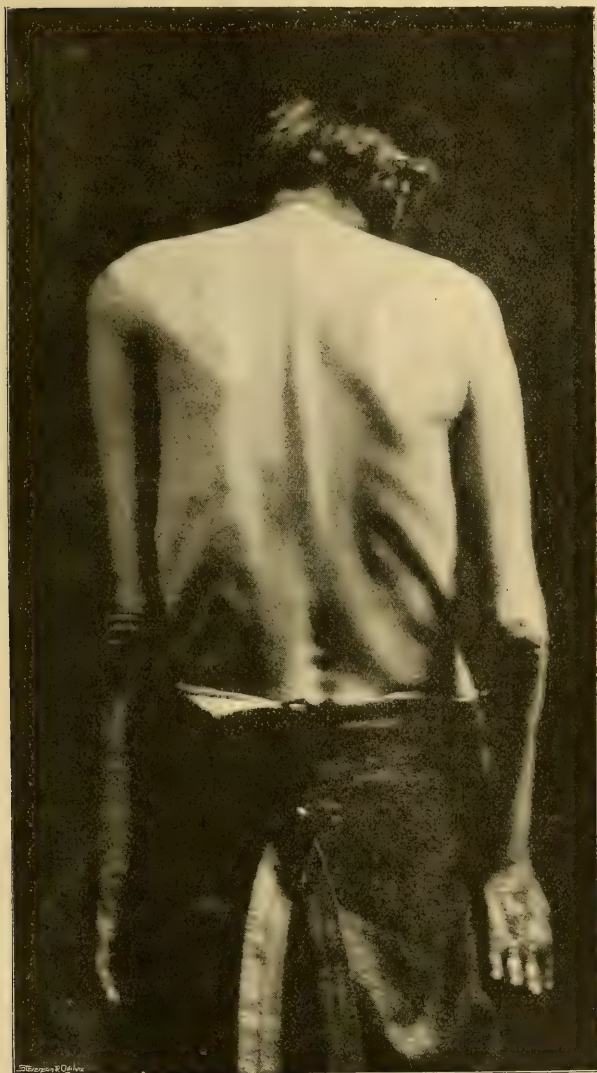


FIG. 56.—*Progressive Muscular Atrophy.*

A deep depression, due to atrophy of the muscles of the back, is seen on each side of the spinal column; the ribs stand prominently out in consequence of atrophy of the intercostal muscles, lower part of the trapezius, and latissimus dorsi.

It is important to note that the atrophy not only involves the muscles, but also seems to affect the subcutaneous and fatty tissues; in advanced stages of progressive muscular atrophy the bones stand out like the bones of a skeleton (see figs. 55 and 56): and the skin of the palms presents a wrinkled appearance.

The temperature of the wasted and atrophied limbs is usually considerably below the normal average.

In the earlier stages, the affected muscles are flaccid, more or less wasted, and weak in proportion to the degree of atrophy which is present. In the advanced stages, the muscular tissue may have almost entirely disappeared. Pressure over the affected muscles does not cause pain. This is an important diagnostic point, for in some forms of peripheral neuritis in which muscular wasting is a prominent symptom—notably in alcoholic peripheral neuritis—muscular tenderness on pressure is a highly characteristic symptom.

Fibrillary twitchings or tremors, which consist in a momentary contraction of individual muscular fasciculi, can usually be seen in the muscles which are undergoing the atrophic process; they are, in fact, more frequent in progressive muscular atrophy than in any other disease. The skin covering the muscular fibre is suddenly raised and stretched as if a thread were made tense beneath it. The patient may be conscious of a momentary quivering sensation in the affected part. I shall have more to say about these fibrillary tremors when I come to speak of the diagnosis.

The electrical condition of the affected muscles varies considerably in different cases. This is apparently due to the fact that the rapidity with which the atrophy and the nerve degeneration are developed, varies in different cases. In the early stages a 'simple diminution' to both forms of current may be the only apparent change: but in most cases, when the disease is well marked, the anodal closing contraction is equal to, or greater than, the cathodal closing contraction. There is, in short, an imperfect form of the reaction of degeneration (see fig. 34). After the detailed consideration which we have already given to the reaction of degeneration, you will easily understand the nature of this change. You must remember that in progressive muscular atrophy some of the motor nerves and motor nerve-endings are degenerated and destroyed, while others are healthy.

Hence, when you apply either the interrupted or the continuous current to the partly atrophied and degenerated muscles and nerves you get contractions; for the healthy or comparatively healthy fibres which persist are capable of contracting to either form of current: but the contractions are less powerful, less forcible, than in health, for many of the muscular fibres are atrophied.

But further, the constant current when applied directly to the atrophying muscles, stimulates not only the healthy motor nerve-endings, but the muscular (sarcous) substance of the muscular fibres which are partly, but not as yet completely, destroyed. The result is that the anodal closing contraction is as readily, or even more readily, produced than the cathodal closing contraction. In the more advanced stages of the disease, when the muscular fibres are entirely destroyed, it may be impossible to elicit muscular contractions with either form of current: but this is rarely the case, for some muscular fibres usually remain even in those muscles which appear to be completely wasted, to the naked eye. As a rule, the faradic irritability disappears before the galvanic irritability; and the electrical excitability of the nerves is, it is said, in some cases retained longer than the electrical excitability of the muscles.

The presence of fibrillary twitchings and of this imperfect form of the reaction of degeneration are most important features from a diagnostic point of view. Fibrillary twitchings are very rarely indeed observed, and the reaction of degeneration is never met with, in the myopathic forms of muscular atrophy.

In some cases of progressive muscular atrophy the mechanical and electrical excitability of some of the affected (atrophying) muscular fibres seems to be for a time increased. There seems, in short, to be a condition of irritable weakness. The affected muscles are more easily excited but at the same time more easily fatigued and exhausted, both by voluntary and electrical stimuli, than in health.

The condition of the reflexes is also variable. When all the nerve cells in any segment are destroyed, the reflexes passing through the segment will of course be completely abolished. But the destruction of the nerve cells is rarely complete, and in the early stages of the disease many nerve cells still remain. Hence, in the early periods of the disease the reflex impulses can still be elicited.

Suppose, for example, that one-third of the nerve cells in the affected part of the anterior horn of the spinal cord was degenerated, and one-third of the muscular fibres connected with the affected portion of grey matter consequently atrophied. In such a case, the reflex impulses would of course be unable to pass through the nerve cells which were degenerated, but could still pass through the nerve cells which remained healthy (see fig. 44). But, as a matter of fact, the reflex movements are in many cases more interfered with than this statement would imply.

Further, you must of course remember, that it is only the reflexes which pass through the affected segments that are interfered with. Since the lesion does not as a rule involve the lumbar enlargement, the reflex movements in the lower limbs are not interrupted; in fact, in many cases they are exaggerated, for in many cases the crossed pyramidal tracts are sclerosed. It was in order that you might understand the rationale of this exaggeration that I deferred the consideration of progressive muscular atrophy until we had considered primary sclerosis of the crossed pyramidal tracts. From these statements you will be prepared to hear, that in many cases of progressive muscular atrophy, the knee-jerks are exaggerated and ankle-clonus is present. When the sclerosis of the crossed pyramidal tracts is very marked—and these are the cases to which Charcot applied the term amyotrophic lateral sclerosis—a condition of complete spastic paraplegia may ultimately be developed in the lower extremities, while the muscles of the upper limbs are in a condition of combined rigidity and atrophy.

In typical and uncomplicated cases of progressive muscular atrophy, the sensory functions are practically intact. In some cases, the patient complains of numbness, deadness, or pins and needles in the affected part; but there is rarely any marked and definite anæsthesia, or any distinct impairment of the sensibility to heat or cold, or to pain.

The functions of the bladder and rectum are not interfered with. Of course, in the later stages of the disease, if the patient should become completely crippled and confined to bed (and this is the usual result in amyotrophic lateral sclerosis but a rare result in the ordinary variety of the disease), the bladder may become paralysed and bed-sores may be developed, as in any other advanced cord lesion.

The general health usually remains good throughout the whole course of the disease. As I have already told you, the fatty tissues of the body seem to waste and disappear, and the patients are usually thin and more or less emaciated. The affected limbs are usually colder than normal. Sugar is occasionally present in the urine; this is, however, quite exceptional: the presence of glycosuria is suggestive of a lesion in the medulla oblongata; in cases of progressive muscular atrophy the presence of sugar in the urine is consequently a very unfavourable symptom.

Course.—The course of progressive muscular atrophy is usually very chronic; the disease generally lasts for years; but this is not invariable. Cases which run a subacute course are occasionally met with; cases have been recorded in which the total duration of the disease seems to have been less than eighteen months; but such cases are exceedingly rare.

Although the usual course of the disease is to advance and progress, the lesion not unfrequently seems to become arrested, more particularly after it has advanced up to a certain point. In some cases, death is the result of extension of the lesion to the medulla oblongata and involvement of the cardiac and respiratory centres. In many cases, the immediate cause of death is an attack of bronchitis. A trivial cold or slight bronchitis, which in an ordinary individual would cause little or no inconvenience, may easily prove fatal in those cases of progressive muscular atrophy in which the intercostal muscles or the diaphragm are paralysed and atrophied. In some cases, complications on the part of the bladder arise in the later periods, but this is not common. Death is sometimes due to the development of some intercurrent disease or complication, which does not directly depend upon the cord lesion.

LECTURE XII

PROGRESSIVE MUSCULAR ATROPHY (*Continued*)

Diagnosis.—In typical and well developed cases, the diagnosis is easy. The recognition of progressive muscular atrophy must be based, partly upon the presence of certain (the positive) symptoms, viz., slowly developing muscular weakness and atrophy, the muscular weakness being in most cases directly proportionate to the muscular wasting; and partly upon the absence of other (the negative) symptoms, viz., the absence of well marked sensory disturbances and of any affection of the bladder or rectum.

The positive and negative symptoms and the very slow and gradual manner in which the disease is developed clearly show that there is a chronic lesion in the lower segment of the neuro-motor nerve apparatus or in the muscles themselves.

All acute lesions in this part of the neuro-motor apparatus (such, for example, as poliomyelitis anterior acuta and the more typical and rapidly developed forms of peripheral neuritis) can be at once excluded.

But in some cases of peripheral neuritis the onset is sub-acute, and in rare cases of progressive muscular atrophy, the disease is developed more rapidly than is usually the case. In such cases, the diagnosis may be attended with more difficulty.

The differential diagnosis of progressive muscular atrophy and of peripheral neuritis.—Inflammation of the peripheral nerves has, during the past few years, assumed a very important place in clinical medicine. We now know that peripheral neuritis is a common condition, and that it may be due to quite a great number of different causes. Now, paralysis and muscular atrophy are conspicuous results of peripheral neuritis, and in some cases, as for instance in alcoholic peripheral neuritis, the atrophy is widely distributed and may involve the small muscles

of the hand which are so constantly affected in progressive muscular atrophy.

But notwithstanding these points of resemblance, the two conditions are readily distinguished. The mode of development and distribution of the paralysis and atrophy are essentially different in the two cases.

In most cases of peripheral neuritis, the onset is more rapid than in progressive muscular atrophy; the paralysis is the first event, and the muscular atrophy is secondary to it; the paralysis is more widely spread and (in the alcoholic form, and this is the most common variety and the one which is most likely to be mistaken for progressive muscular atrophy) the muscles of the leg and foot are more markedly affected, or at least as much affected, as those of the forearm and hand. Further, in peripheral neuritis sensory symptoms (anæsthesia and hyperæsthesia) are usually prominent features in the clinical picture. A definite cause, such as alcohol, can usually be elicited. The inflamed nerve trunks are tender on pressure, and in the alcoholic form—and this is a very characteristic symptom—muscular tenderness on pressure is usually well marked. Lastly, the paralysis and atrophy which result from peripheral neuritis tend to get well, and are in many cases completely recovered from, though recovery may be very slow (several months or even longer in bad cases): while in progressive muscular atrophy the muscular weakness and atrophy tend to increase and extend.

The differential diagnosis of the spinal (Aran-Duchenne) type and of the idiopathic (myopathic) forms of progressive muscular atrophy will be better understood after I have described pseudo-hypertrophic paralysis and the allied forms of idiopathic muscular atrophy.

The differential diagnosis of the typical spinal (Aran-Duchenne) type of progressive muscular atrophy and of the peroneal type of progressive muscular atrophy (which is probably myelopathic rather than myopathic in nature) will also be subsequently considered.

The chief difficulty in diagnosis occurs in the earlier stages when the muscular wasting is localised and limited to the small muscles of the hand and thumb. But even in these cases, a careful observer is usually able to come to a correct conclusion as to the nature of the case.

In cases of ulnar nerve paralysis, leprosy, amyotrophic lateral sclerosis, pachymeningitis cervicalis hypertrophica, tumors pressing upon the spinal cord and anterior nerve roots at the lower end of the cervical enlargement, and syringomyelia, the small muscles of the fingers and thumb may become atrophied, and a condition of the hand which more or less closely resembles that characteristic of progressive muscular atrophy may be produced. But in each and every one of these diseases there are associated symptoms which unmistakably show that the muscular wasting is not due to progressive muscular atrophy.

The differential diagnosis of progressive muscular atrophy in an early stage and paralysis due to a lesion of the ulnar nerve. In progressive muscular atrophy, all the fingers are equally affected; but in the clawed hand due to a lesion of the ulnar nerve, the little and ring fingers are much more bent than the middle and forefinger, for the first and second interossei muscles, which are supplied by the median nerve, escape (see figs. 48 and 49). In progressive muscular atrophy, sensation is not affected; but in ulnar nerve paralysis, there is anæsthesia on both sides of the little and on the ulnar side of the ring finger. In progressive muscular atrophy, there are no trophic alterations in the skin and nails; but in ulnar paralysis, trophic alterations in the skin and nails of the little finger are often present. Progressive muscular atrophy is slowly, while ulnar paralysis is usually rapidly developed. In ulnar paralysis, a history of injury to the ulnar nerve may be forthcoming, or a wound in the course of the nerve may be present.

In cases of bilateral paralysis of the ulnar nerve, the physician is apt to be thrown off his guard and to suppose that the case is one of progressive muscular atrophy, for bilateral paralysis of the ulnar nerve is very rare. A few years ago a case of this kind came under my own observation. Both hands presented the bird-claw condition; at first sight the case looked like one of progressive muscular atrophy. But on closer inspection the true nature of the case was apparent. The little and ring fingers were much more bent than the middle and forefingers; the characteristic anæsthesia was present; and on inquiry a history of injury was forthcoming. The patient had slipped and fallen on both elbows; a bilateral injury to the ulnar nerves had resulted. An interesting point in the case was the fact that the

patient was the subject of urticaria scripta. I have noticed a tendency to inflammation of the peripheral nerves in other cases of urticaria scripta.

Leprosy.—In some other affections, the small muscles of the hand may become atrophied and wasted. Leprosy, a disease which is rarely seen in this country, is one of them. In anæsthetic leprosy, as in progressive muscular atrophy, the muscular atrophy and weakness are slowly and gradually developed, but the diagnosis presents no difficulty. In anæsthetic leprosy, the ulnar and other nerves in the forearm or upper arm are enlarged; in many cases, they can be felt as hard rigid cords, and they can in some cases be seen standing out in the forearm. Other symptoms and signs of anæsthetic leprosy (such as anæsthesia in patches, skin eruption, glandular enlargement in the groin, etc.) will probably be present. Further, the history (the locality in which the patient has lived and contracted the disease) affords corroborative evidence of the true nature of the case.

Amyotrophic lateral sclerosis.—This condition, which is now thought by some authorities to be merely a variety of progressive muscular atrophy, is easily distinguished from the ordinary form of progressive muscular atrophy. I will presently refer to its characteristic features in detail. The spastic condition in the lower extremities; increase of the deep reflexes both in the lower and upper limbs clearly show that the pyramidal tracts are involved in a marked degree, and differentiate the condition from the ordinary form of progressive muscular atrophy.

Pachymeningitis cervicalis hypertrophica, and tumors which press upon the spinal cord in the lower cervical region.—These conditions are more likely to be confounded with amyotrophic lateral sclerosis than with the ordinary (uncomplicated) form of progressive muscular atrophy, from which they are readily distinguished. You will better appreciate the points of distinction after I have described spinal meningitis. All I need now say is that in both affections (pachymeningitis cervicalis hypertrophica and tumors pressing upon the cervical region of the cord) symptoms indicative of implication of the posterior roots (pain and anæsthesia in the upper extremities) are usually prominent; and symptoms due to pressure upon the cord itself (weakness, paralysis, rigidity, spasm, exaggeration of the deep reflexes in the lower extremities) are often present. The wasting of the

small muscles of the hand and the *main en griffe* (which may be present and which may suggest progressive muscular atrophy) are not the only symptoms—unless indeed the anterior nerve roots in the lower cervical region are alone pressed upon and involved; but such limitation is very rare, if it ever occurs.

Syringomyelia.—In this condition, in which a cavity is developed in the centre of the cord, localised muscular atrophy is of frequent occurrence, for the region of the anterior horn is often invaded by the lesion. When the anterior cornu in the cervical region is implicated, the muscular wasting may closely simulate that due to progressive muscular atrophy. But in syringomyelia, other characteristic symptoms, notably loss of the temperature-sensibility, and trophic lesions in the skin, nails or bones, are almost always present. Further, the distribution of the muscular wasting and the order of its development and spread are usually different from that which is present in progressive muscular atrophy.

Lead Poisoning.—In this condition, the cause of the muscular atrophy is usually clear enough. A distinct history of exposure to lead is usually forthcoming, and other symptoms characteristic of lead impregnation (blue line on the gums, colic, anæmia, etc.) are almost always present. Further, the muscular atrophy is usually different in the two cases; and in doubtful cases the practitioner can always fall back upon the therapeutic test—the effect of treatment. I have, however, seen at least one case of undoubted progressive muscular atrophy in which the patient had suffered from lead poisoning, and in which the plumbism was perhaps a cause (or possibly the cause) of the disease.

Localised lesions of the anterior horn of grey matter are occasionally developed in the course of locomotor ataxia, primary spastic paraplegia, and other chronic diseases of the spinal cord. But there is no difficulty in distinguishing the localised muscular atrophy which is produced in this way from that which is due to progressive muscular atrophy. In the great majority of cases, the muscular wasting is localised and limited to the lower extremities; further, it shows little or no tendency to progress and involve other muscles. This is a most important diagnostic point, for *the* great feature of progressive muscular atrophy is the tendency to extend and to involve one muscle after another.

The diagnostic significance of fibrillary twitchings and tremors.

Before leaving the subject of diagnosis, let me refer to the significance of fibrillary twitchings. I have already told you that although fibrillary twitchings and tremors are more frequent in progressive muscular atrophy than in any other disease, they are in no way pathognomonic or distinctive of that condition. In fact, quite the contrary. Fibrillary twitchings are by no means uncommon in neurasthenia and other functional conditions; they also occur in many organic affections of the spinal cord in which a subacute or chronic lesion involving the anterior cornual region is attended with muscular weakness, paralysis and atrophy—in traumatic myelitis (during the stage of recovery) and arthritic muscular atrophy, for example.

So far as my observation enables me to judge, fibrillary twitchings may result either from a condition of irritable weakness of the nerve cells—a functional condition—or from slow degeneration and destruction of the nerve cells—an organic condition.

Almost every one, I suppose, has experienced fibrillary twitchings in the eyelid; they are common enough, too, in other parts of the body. A layman who is affected with these fibrillary twitchings, and who knows nothing about progressive muscular atrophy, is rather amused than otherwise by them; but a doctor who observes these twitchings in his own person sometimes at once jumps to the conclusion (it is a big jump, but he makes it) that he is going in for progressive muscular atrophy. The mental anxiety and depression which result from this idea are often very great. As a consequence, the general health, which was previously perhaps below par, becomes still further deteriorated. This imaginary form of progressive muscular atrophy is a doctor's disease. It is one of the imaginary diseases with which doctors and medical students are apt to be affected. A very considerable percentage of medical students imagine, as many of you doubtless know from personal experience, that they are the subjects of heart disease. But I need not go into details. We have already considered this subject in connection with the valvular lesions of the heart. What I want to impress upon you now is this, that the mere presence of fibrillary twitchings is no evidence of progressive muscular atrophy or indeed of any other serious disease of the nervous system. In the diagnosis of progressive muscular atrophy, and indeed of almost every other disease, the whole

circumstances and facts of the case must be taken into account. In making a diagnosis, you must carefully avoid attaching too much importance to any one individual fact or symptom. Fibrillary twitchings *per se* are of little or no importance. It is very different if the fibrillary twitchings are associated with well marked and localised muscular atrophy.

Do not however forget the point which I have previously emphasised that fibrillary tremors which are in most cases such a conspicuous feature of the Aran-Duchenne form of progressive muscular atrophy are very rarely indeed met with in the idiopathic (myopathic) forms of the disease. What I mean to say is this, that given a case of progressive muscular atrophy, the presence of fibrillary twitchings is an important diagnostic sign of the myelopathic as distinct from the myopathic form of the disease.

Prognosis.—Progressive muscular atrophy usually runs a very chronic course; but in some cases the duration of the disease is comparatively short; cases have been recorded in which the disease terminated within eighteen months from its commencement. Such a rapid course is, however, quite exceptional. The prognosis as regards duration is consequently, in most cases, fairly good.

The prognosis as regards complete recovery is very bad. The fact that the lesion is degenerative in character shows that there is an inherent tendency to decay in the affected nerve elements. The degenerative atrophy destroys the nerve cells, and so far as we know, nerve cells which have once been destroyed are never restored.

The prognosis as regards arrest is uncertain, and in most cases unfavourable. It is perfectly true that the morbid process is sometimes arrested; but in the majority of cases, the degeneration extends and gradually goes from bad to worse. In most cases, although temporary periods of apparent arrest may occur, there is no real arrest—simply a period of temporary quiescence. In other cases, after the lesion has advanced to a certain stage, a true arrest seems to occur. In the case which is represented in figs. 55 and 56, the patient lived for several years (the disease being in a static or arrested condition) after almost all of the muscles of the upper extremities and many muscles of the

back and thorax had been completely destroyed. Arrest in the early stages (when, for instance, the atrophy is limited to the hand muscles) is in my experience very rare. Gowers thinks that arrest is more likely to occur in those cases in which the corresponding muscles on opposite sides of the body are simultaneously or almost simultaneously affected, than in those cases in which the atrophy is irregularly developed.

These are general statements. They are of great importance as general guides to prognosis. But in practice we have to deal with individual patients. When a patient comes before us with progressive muscular atrophy, we have to endeavour to determine what the course of the disease is likely to be in his particular case. It is a matter of comparatively little importance to the patient what the result is in the average run of cases; what he wants to know is what course the disease will be likely to take in his particular case. It is small comfort to him to know that according to the books the disease runs a progressive course from bad to worse, in eighty per cent. of the cases, and that in the remaining twenty per cent. it becomes arrested. What he naturally wants to know is whether the disease is likely to be arrested in his individual case.

In trying to form an opinion on this point, the following are the more important points which have to be taken into account:—The length of time which the disease has existed; the rapidity with which it appears to be progressing; the number of muscles which are affected; the degree of atrophy in the muscles which are affected; whether the respiratory muscles are involved or not; the presence or absence of bulbar symptoms; the age of the patient; the circumstances and surroundings of the patient; and the presence or absence of complications (phthisis, kidney disease, etc.).

You would naturally expect that the disease would be less amenable to treatment in old than in young people; and speaking generally this is the case. Degenerative processes are more common (and after they have commenced perhaps more apt to progress) in old than in young people; but the age of the patient is a very uncertain guide, for the mere fact that progressive muscular atrophy (a disease which is due to a degenerative process) occurs in a young person, shows that the normal vitality, so to speak, which the nerve cells ought to possess in youth is not present.

When arrest does take place, it very often occurs, as I have already stated, after a large number of muscles, perhaps all of the muscles of both upper extremities, have become atrophied: in other words, after the majority of the motor nerve cells in the cervical enlargement of the cord are destroyed.

In every case of progressive muscular atrophy, symptoms indicative of involvement of the nerve nuclei in the medulla oblongata should be carefully looked for; bulbar symptoms are always of very grave significance. The presence of sugar in the urine, even without any other bulbar symptoms, is unfavourable. In those cases in which bulbar symptoms are developed early, the course is usually very rapid; fortunately in most cases bulbar symptoms are not developed until the terminal stages of the disease.

When the intercostal muscles or the diaphragm are affected, even in a slight degree, the prognosis is bad; for in these cases a slight bronchitis may, in consequence of the difficulty in getting rid of the secretions which accumulate in the bronchi, prove fatal.

Paralysis of the bladder is another very unfavourable condition; fortunately it is very seldom developed until the terminal stages of the disease; and even then it is rare.

In estimating the prognosis, the presence or absence of associated lesions in other parts of the cord must of course be taken into account; in amyotrophic lateral sclerosis, for example, the prognosis is more unfavourable, both in respect to duration and probable arrest, than in the ordinary form of the disease in which the anterior cornual region is alone involved.

It is perhaps hardly necessary to say that the financial circumstances and surroundings of the patient materially modify the prognosis. One essential point in the treatment of the disease is to protect the patient from attacks of bronchitis and other pulmonary complications such as phthisis. Patients who are well off, and who are consequently well fed, well housed, well clothed, and well looked after generally, are, other things being equal, likely to survive longer than others who are less favourably situated in these respects.

Treatment.—Almost all observers are agreed that the treatment of progressive muscular atrophy is very unsatisfactory; this

is only what we would expect since the morbid process is a degeneration which tends in most cases to progress from bad to worse. Gowers seems to differ from this view. In the last edition of his work on the spinal cord he makes the remarkable statement that 'in seven almost consecutive cases in middle life, the treatment (hypodermic injections of strychnine) has been followed by arrest within a month of its commencement, and the arrest has been permanent in all cases but one. In the senile cases,' he says, 'the treatment has failed, but in most of them the disease was in an advanced stage, and the lumbar cord had begun to suffer.'¹ In some of the cases in which the result was prompt and distinct, strychnia given by the mouth had failed. He recommends that one injection should be given daily, at any convenient place. He gives one-hundredth of a grain of the nitrate of strychnia at first, quickly increased to one-fortieth of a grain. It will be very remarkable if future observations confirm these results.

Arsenic is another remedy which is of undoubted value in degenerative nerve processes; I am satisfied that it is beneficial in some cases of progressive muscular atrophy. Nitrate of silver is probably also useful. Quinine is probably also beneficial as a general tonic. So far as I know, these are the only drug remedies which are likely to have a beneficial influence upon the course of the disease; and so far as my experience enables me to judge—and this I fancy is the experience of almost every physician—their influence for good is very small, at all events when given by the mouth. In most of the cases in which the disease becomes arrested, the arrest is probably due to a natural process rather than to the influence of drugs. It is impossible I think to predict in any individual case whether an arrest will take place or not.

In treating cases of progressive muscular atrophy, it is essential to attend to the condition of the general health, and to avoid everything which is likely to produce depression or to hasten the development of the degenerative process. The patient should be well fed, well housed, well clothed and carefully protected from cold and all depressing conditions, such as mental or emotional excitement. It is especially important to avoid all causes of muscular fatigue. A certain amount of muscular

¹ *Diseases of the Nervous System*, vol. i. p. 497.

exercise is beneficial, for it is important to maintain the functional activity and healthy nutritive condition of the nerve cells and muscles which are as yet unaffected; but exercise which causes fatigue and throws a strain upon the nerve cells and muscles which are in process of degeneration should be avoided.

Fresh air and sunshine are eminently desirable; consequently, patients who are well off should be advised to spend the winter and early spring in the sunny South. Egypt, the Riviera, the Canaries are perhaps the most suitable climates.

Local electricity and massage are, I think, useful; but they must be carefully and judiciously employed. Powerful electric currents are likely to do harm rather than good, and rough massage is injurious. The form of current (whether the faradic or galvanic) is not perhaps a matter of much importance. A current just sufficiently strong to produce muscular contractions is all that is necessary. I usually recommend a weak faradic current. I believe that local electricity, gentle massage, and muscular exercise, if judiciously and cautiously employed, are undoubtedly beneficial in some cases. They help, I think, to maintain the nutritive condition of the healthy (unaffected) muscular fibres and perhaps prevent the degenerative (atrophic) changes advancing as rapidly as they might otherwise do in the affected muscular fibres. But further, these means of local treatment encourage the patient to hope. He sees, feels, and realises that something definite and positive is being done, day by day, to prevent the development of the disease. He is thereby enabled to maintain a cheerful and to some extent at least, a hopeful tone of mind; and this of itself undoubtedly excites a beneficial influence upon the progress of the lesion. Or rather perhaps I should say that if the patient becomes despondent and gives up all hope, the progress of the disease is apt to be injuriously affected by the state of mental depression. In this, as in many other diseases, mental therapeutics play a most important part in treatment. Speaking generally, a cheerful and hopeful condition of mind has a tonic and invigorating action upon the nutritive processes of the body; while a despondent and hopeless frame of mind has a most depressing and injurious influence upon these nutritive changes, and is apt, I think, to favour the development and to accelerate the course of many morbid processes. Do not misunderstand me on this

point. I do not mean to say that you should tell a patient who is suffering from progressive muscular atrophy that local electricity and massage, or indeed any other form of treatment, will cure him. That would be nothing less than arrant quackery. But, believing as I do that these remedies are in some cases beneficial in preventing the rapid development of the disease, I maintain that they should be employed, and that the patient should be encouraged to hope that in his case they may perhaps be, in some degree at least, beneficial.

The catalytic action of the constant current applied to the spine is possibly also of some use; but, as I have more than once told you, it is very difficult to draw correct conclusions upon this point; and I speak with great reserve regarding it. The position which I take up is that a weak constant current applied to the spine cannot possibly do any harm, and that although I am not personally convinced that it actually does good, I do not feel justified in saying that it never does any good. It is possible, I think, that it may in some cases be beneficial. Hence it is, I think, advisable to give the patient the benefit of the doubt. But I repeat that if the constant current is applied to the spine, a weak current only should be employed.

Throughout the course of the disease, and especially in those cases in which the respiratory muscles are involved, it is of the greatest importance to avoid everything which is likely to produce bronchial catarrh or any other respiratory complication. When, too, the bladder is paralysed, great care must be taken to prevent the development of septic cystitis. I have more than once told you that one of the great dangers in diseases of the spinal cord is the development of bladder and kidney complications. Fortunately in most cases of progressive muscular atrophy the risk of bladder and kidney complications is not great. But it is essential to remember that they do sometimes occur.

During the last stages of the case, when the patient is confined to bed, care must of course be taken to prevent the development of bed-sores. The risk of bed-sores is greater in amyotrophic lateral sclerosis than in the ordinary form of progressive muscular atrophy.

In the later stages of uncomplicated cases of progressive muscular atrophy the duration depends in great part upon the way in which the patient is looked after and nursed.

In those cases in which there is a definite history of previous syphilis, iodide of potassium and mercury should of course be tried.

AMYOTROPHIC LATERAL SCLEROSIS

In our study of the Aran-Duchenne type of progressive muscular atrophy we have seen that the crossed pyramidal tracts in the lateral column of the spinal cord are not unfrequently sclerosed; and that some authorities believe that there is no real distinction between progressive muscular atrophy on the one hand and amyotrophic lateral sclerosis on the other. The correctness of this view is, I think, questionable. I doubt whether the cases to which Charcot applied the term '*sclérose latérale amyotrophique*' are mere modifications of the ordinary (spinal) form of progressive muscular atrophy.

Morbid Anatomy.—In amyotrophic lateral sclerosis the lesion consists of a sclerosis of the crossed pyramidal tracts and an atrophic destruction or degeneration of the multipolar nerve cells in the anterior cornua of the spinal cord.

The sclerosis of the crossed pyramidal tracts is, according to Charcot, more extensive than the sclerosis (secondary descending degeneration) which follows a cerebral lesion. The lesion in the anterior cornua seems identical with, or at all events very similar to, that of progressive muscular atrophy. According to Charcot the lesion in the anterior cornua is secondary to the lesion in the crossed pyramidal tracts.

In some cases (whether this is a frequent or a constant condition it remains to be shown), the pyramidal fibres in the crus cerebri and internal capsule are also affected. Morbid changes in the motor cortex of the brain (degeneration and atrophy of the large motor nerve cells) have also been described. These facts suggest that in amyotrophic lateral sclerosis the disease has its starting-point in the motor cerebral cortex.

Clinical History.—The chief symptoms which characterise amyotrophic lateral sclerosis are:—loss of muscular power, muscular atrophy, rigidity and tension of the muscles, and exaggeration of the deep reflexes.

Charcot described three stages of the disease.

In the first stage, which usually lasts from four to twelve months, the symptoms are, for the most part, confined to the upper extremities. The first symptom which is developed is muscular weakness; it is soon followed by atrophy which is more diffuse than that which characterises the Aran-Duchenne type of progressive muscular atrophy, and by rigidity and contractures. The French school of neurologists lay great stress upon the fact that the muscular weakness (paresis or paralysis) precedes the muscular atrophy—the very reverse of the condition which obtains in the Aran-Duchenne type of progressive muscular atrophy.

Fibrillary twitchings can almost always be observed in the

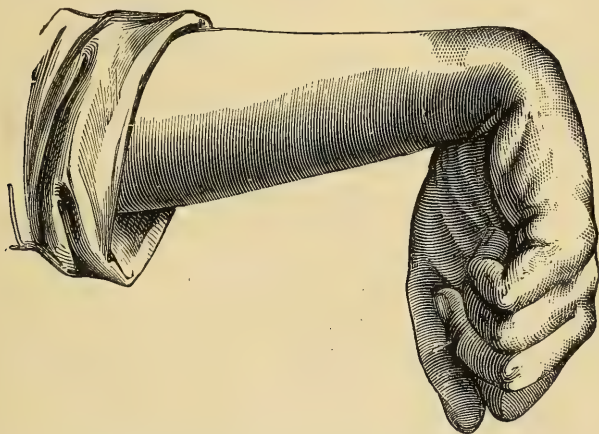


FIG. 57.—*Position of the hand in amyotrophic lateral sclerosis.*—(After Charcot.)

affected muscles, and the reaction of degeneration is usually developed in the later stages of the disease.

As the result of the muscular tension and contractures, deformities are produced, and the hand often becomes fixed in the position shown in fig. 57. According to Grasset and Rauzier, the position and condition of the upper extremities is highly characteristic. 'The arm is kept applied to the side of the body, and the muscles of the shoulder offer resistance when one attempts to separate it from the trunk. The forearm is semiflexed and pronated; passive movements made with the object of counteracting this deformity cause pain. The thumb is flexed, and the fingers are forcibly flexed into the palm.'¹

¹ *Maladies du Système Nerveux*, page 660.

The muscles of the neck are in some cases affected, and the head may be rigidly fixed in one position by the spasm and contracture which are present.

During this, the first, stage of the disease, the deep reflexes both in the upper and lower extremities are exaggerated. A chin-jerk (jaw-jerk) can in some cases be elicited.

In the second stage, the lower extremities become invaded; the muscles of the legs become rigid, the knee-jerks are greatly exaggerated, ankle-clonus is present; in short, the condition of the lower limbs is identical with that which results from primary sclerosis of the crossed pyramidal tracts. There is no marked disturbance of sensation and no affection of the bladder or rectum.

After this condition of spastic paraplegia has lasted for some time, the morbid process may extend to the anterior cornua in the lumbar enlargement; the muscles of the lower extremity then become atrophied, and the rigidity, spasm and exaggeration of the deep reflexes decrease.

During this, the second, stage of the disease, the atrophy in the upper extremities continues to progress and the rigidity and contracture to diminish.

In the third stage, the paralysis and muscular atrophy are exaggerated and the motor nerve cells in the medulla oblongata are affected; bulbar symptoms are then developed, and death results.

In exceptional cases, the motor nerve cells of the medulla oblongata are affected before those of the spinal cord, and the disease appears to extend downwards. Occasionally, the lower extremities are first affected; the morbid process then appears to extend upwards to the cervical region of the cord.

The clinical features which I have just described apply to typical cases. It is important to remember that many cases are not typical. Cases which seem to form intermediate types between the Aran-Duchenne form of progressive muscular atrophy and amyotrophic lateral sclerosis are, comparatively speaking, common. This seems to me to be the chief argument in favour of the view that the two diseases are mere modifications of one and the same morbid process.

Course.—So far as is known, the disease is invariably fatal. The duration seems to vary in different cases, but death usually

results in from one to three years from the commencement. In some cases the course of the disease is measured by months rather than by years. The rapidity with which the disease progresses is an important point of distinction between amyotrophic lateral sclerosis and the ordinary (Aran-Duchenne) type of progressive muscular atrophy.

Charcot's observations would seem to show that the disease is more common in females than in males; but further information is required on this point. Most of the cases which I have seen have been in males. The disease usually develops between the ages of 20 and 50.

The causes of the condition are unknown.

Diagnosis.—The mode of development, the character of the symptoms and the course which they pursue, distinguish the condition, clinically at all events, from all other diseases of the spinal cord.

The chief points of importance from a diagnostic point of view are:—the muscular paralysis and atrophy, the rigidity and muscular tension, the exaggeration of the deep reflexes, and the absence of any characteristic disturbance of sensation and of the functions of the bladder and rectum.

The differential diagnosis of amyotrophic lateral sclerosis and of progressive muscular atrophy.—As I have already told you, some observers are of opinion that the Aran-Duchenne form of progressive muscular atrophy and amyotrophic lateral sclerosis depend essentially upon one and the same lesion. While prepared to grant that intermediate cases, which seem to form connecting-links between the two distinct types, are by no means rare, I doubt the correctness of this view. As I have already pointed out, in many cases of typical progressive muscular atrophy there is, so far as can be judged from the clinical symptoms, no sclerosis of the crossed pyramidal tracts; and the correctness of this opinion is in some cases confirmed on post-mortem examination.

Further, the mode of distribution and development of the muscular atrophy are different in the two cases. In typical cases of amyotrophic lateral sclerosis, the muscular atrophy is less localised (more diffuse) and is developed more rapidly than in typical cases of progressive muscular atrophy; and in many

cases, as I have already pointed out, it appears to be preceded rather than followed by muscular paralysis. Although in both diseases the lesion usually commences in, and is chiefly confined to, the cervical enlargement of the cord, the exact point of departure is usually different; in typical cases of progressive muscular atrophy, the small muscles of the hand are first affected; while in amyotrophic lateral sclerosis, the muscles supplied by the musculo-spinal nerve are often first involved.

Again, although the deep reflexes are often exaggerated in progressive muscular atrophy, in that disease there is no muscular rigidity and no contracture.

Further, amyotrophic lateral sclerosis usually runs a more acute course than progressive muscular atrophy.

The differential diagnosis (1) of amyotrophic lateral sclerosis and of pachymeningitis cervicalis hypertrophica and (2) of amyotrophic lateral sclerosis and of extra-medullary tumours involving the upper part of the cervical enlargement will be more appropriately considered afterwards.

Prognosis.—This is most unfavourable.

Treatment.—All the therapeutic measures which have hitherto been employed seem to be useless and inadequate to check the progress of the affection.

Counter-irritation, more particularly the application of the actual cautery over the cervical enlargement, and the internal and local means of treatment which have been recommended for the treatment of progressive muscular atrophy should be employed.

LECTURE XIII

THE PROGRESSIVE MUSCULAR DYSTROPHIES¹

IN the last lecture, Gentlemen, we finished the consideration of the spinal form (the Aran-Duchenne type, as it has been termed) of progressive muscular atrophy. We saw that it is due to a slow and gradual destruction of the multipolar nerve cells in the anterior horn of the spinal cord; it is consequently a *myelopathic* muscular atrophy.

To-day I wish to direct your attention to some other forms of muscular atrophy, which, so far as our present knowledge and methods of investigation enable us to judge, are of *myopathic* origin, i.e. due to a lesion in the muscles themselves.

The subject is a difficult and complicated one, for several of the forms of myopathic muscular atrophy to which I am about to refer have only been recently differentiated, and there is still some uncertainty as to their true pathology and as to the relationship which they bear one to another. Further, cases of myopathic atrophy (the pseudo-hypertrophic type alone excepted) are, in this country at all events, rare, and the opportunities of studying them seldom occur.

The most striking clinical feature of the myopathic forms of progressive muscular atrophy, as of the spinal form of progressive muscular atrophy, is a slow, gradual and (usually) progressive muscular wasting, which is manifested externally and perceived by the patient as a gradual and progressive loss of motor power.

Now, muscular atrophy may of course be due to a lesion in any part of the lower division of the neuro-motor nerve apparatus (anterior horn of the spinal cord, anterior nerve-roots, peripheral

¹ Since there is still some doubt as to the ultimate causation of the progressive muscular dystrophies or muscular atrophies of myopathic origin, and since in their clinical features they closely resemble certain forms of muscular atrophy due to diseases of the spinal cord, they are most appropriately considered in connection with the diseases of the spinal cord.

motor nerves, or motor nerve-endings in the muscles), or to a lesion of the muscles themselves; that is to say, to a lesion (1) in the anterior horn of the spinal cord, (2) in the nerve tract which passes from the anterior horn to the muscles, or (3) in the muscles.

We have seen that acute destruction of the multipolar nerve cells in the anterior horn of the spinal cord is followed by rapid and marked muscular atrophy; and you know that acute destruction of a motor peripheral nerve produces the same result.

Further, our study of the Aran-Duchenne type of progressive muscular atrophy and of amyotrophic lateral sclerosis has shown us that slow and gradual destruction of the multipolar nerve cells (the trophic centres) in the spinal cord, is attended with a slow and gradual muscular atrophy. Slow and gradual destruction of the motor fibres of the peripheral nerves will necessarily be followed by the same result. But slow and gradual destruction of the motor nerve fibres without any corresponding affection of the sensory nerve fibres is very rarely met with as a primary condition. Destruction and degeneration of the peripheral nerves, the result of peripheral neuritis, is common enough; but in the great majority of cases, peripheral neuritis runs an acute or subacute course; and it is only in quite exceptional cases of peripheral neuritis that the motor nerve fibres are alone implicated. Except in those cases in which degenerative and atrophic changes in the nerve fibres are secondary to a slow and gradual destruction of the nerve cells in the anterior horn of the spinal cord (their trophic centres), slow and gradual destruction and degeneration of the motor, apart from the sensory, fibres of the peripheral nerves very rarely indeed occurs.

A slowly developed and progressive muscular atrophy, without associated sensory symptoms, is consequently very rarely the result of a primary lesion of the peripheral nerves, that is to say, it is very rarely neuropathic. There is indeed one form of progressive muscular atrophy which is supposed by some authorities to be due to this cause—I refer to the peroneal type of the disease. But whether in that affection the lesion is actually situated in the peripheral nerves is, as yet, by no means certain. I shall refer to this question in more detail presently. (See page 270.)

From these considerations, it will be obvious to you that a slowly developed progressive muscular atrophy, which is entirely

unassociated with sensory symptoms, is in the great majority of cases either myelopathic or myopathic, i.e. due to a lesion in the spinal cord or the muscles themselves. As a matter of fact, most of the forms of progressive muscular atrophy which I propose to bring before your notice in this lecture are, so far as our present methods of investigation enable us to judge, of myopathic origin. This statement is based upon the results of histological examination; for up to the present time no definite and constant lesions have been detected in these cases, either in the spinal cord or in the peripheral nerves. It is true that in individual cases such lesions have been described, but in the great majority of instances which have been carefully examined by competent observers, working with modern methods, the spinal cord and peripheral nerves have been found to be absolutely healthy.

But granting this, it is not impossible to conceive—and the passage which I will presently quote from Erb gives some support to this view—that the *muscular* changes, which, so far as our present means of examination enable us to judge, constitute the primary lesion, may be the result of changes in the trophic centres for the muscles (multipolar nerve cells of the anterior cornua), which are too delicate to be recognised by our present methods of investigation. The healthy functional and structural condition of the muscular fibres of course depends upon, is governed by, the trophic nerve cells. It is not unreasonable, I think, to suppose that an altered condition of these trophic centres—a change which is, as we term it, functional and unattended with any obvious structural modifications, or at all events with any alterations which we can at present recognise—may exist as the peculiar mode of function in certain individuals (the subjects of myopathic atrophy) and may, in them, lead to the altered state of muscular nutrition which is the cause of the atrophy and hypertrophy of the muscular fibre and of the increase of the interstitial fibrous tissue which seems to be the primary pathological condition. The statement of Erb which seems to me to give some support to this view is as follows:—

‘On the other hand, the results of the examination of the nervous system have, up to the present, been almost wholly negative. Leaving out of account Lichtheim’s case and the older cases of pseudo-hypertrophy, we have had recently a series

of exact records from Fr. Schultze, Landouzy, Déjerine, P. Marie, Dreschfeld, Westphal, and Singer, and neither in the spinal cord nor in the peripheral nerves have any noticeable changes been observed. In accordance with these records, we should be entitled to view this dystrophy as a disease limited to the muscle-substance, and as a truly primary myopathy, were it not that some facts have come out which warn us to exercise great caution in this direction. Heubner's case deserves special mention. Though an undoubted case of dystrophy, he found there was extensive atrophy of the large cells in the anterior horns of grey matter. Frohmaier has a similar case with the changes in it less marked. In this new light the more trifling alterations observed by Singer and the older observations in pseudo-hypertrophic cases of L. Clarke, Gowers, Kesteven, Bramwell, and J. Ross, gain a certain significance. But in the meantime we must say this, that in progressive muscular dystrophy in its various forms, the nervous system must be considered to be, as a rule, and for our present methods of investigation, normal.

This is the place in which to say a little about the proper nature, pathogenesis, and exact seat of the lesion. It is not necessary to prove further that it is neither a simple atrophy of the muscles nor an inflammatory affection with its consequences; there is unquestionably a more complicated disturbance of nutrition, regarding the nature of which, for the present, I would rather not express a definite opinion.

Pathological anatomy has led many to consider it simply as a local muscular affection, quite independent of the central nervous system, a pure myopathy. But a good deal of doubt has been expressed (for example, by Knoll) regarding this view, and I myself, in my earlier work, brought forward some reasons which prevented me from accepting it without more inquiry. A much larger experience, a good deal of consideration given to the subject, and most of all the results of the previously mentioned autopsies have confirmed me in my scruples.

The considerations which weigh with me are various. The muscles depend for their nutrition to a very large extent on trophic nerve centres; the localisation of this atrophy frequently follows in a noticeable way the exact course of the nerves in a plexus, or the disposition of the centres in the central organ, and occasionally we find a case of spinal amyotrophy (Strümpell's case) presenting an almost exactly similar arrangement. Hereditary influence is of great importance; mental aberrations are common among the patients, and other neuroses frequently occur in their families. Further, even in the undoubtedly spinal cases, such as acute anterior poliomyelitis, similar morbid changes

(hypertrophy, proliferation of nuclei, division of the fibres) both in the muscles and connective tissue have been pointed out by W. Müller, Déjerine-Huet, Joffroy-Achard, and Hitzig. In pseudo-hypertrophic cases, malformations and changes of a minor kind in the spinal cord have been met with. When I consider these facts, and bear in mind further the results of Heubner's and Frohmaier's observations, I cannot avoid the suspicion that after all the affection may be dependent on the nervous system. It is tempting to suppose, as I formerly expressed it, that we have to do with a kind of trophoneurosis, having its origin in the trophic centres of the cord—a disturbance of the function of these centres which finds its expression in the very complicated muscle changes of the disease. While on this supposition there are, as a rule, no coarse nerve changes, now and then, and after the affection has lasted a long time or been very intense, such a change does become visible.

The idea is inevitable that if something like this is the case the relations between dystrophy and spinal amyotrophy will turn out again in the end to be close and intimate. The latter would represent an affection of the trophic centres that from the very first is a distinct coarse anatomical lesion taking effect in a degenerative atrophy of the muscles with fibrillar twitchings, reaction of degeneration, etc.; the former at the outset would be merely a functional disturbance of these centres, conditioned probably by different causes, and expressing itself as muscular dystrophy with all its characteristic symptoms. At the same time, there would remain the possibility that even this merely functional disturbance might in the long-run become associated with a coarse lesion of the centres. Many things about these affections would agree very well with such a supposition, among them the occasional occurrence of the reaction of degeneration in dystrophic cases and the occasional initial localisation of spinal amyotrophy in the shoulder and trunk. But I will not spin out this discussion to any greater length; the whole question is by no means yet ready for decision, and the future alone can lift the veil and reveal the finer processes that as yet lie hid from us.¹

The chief facts in favour of the myopathic origin of the lesion are:—(1) the absence of any definite and constant changes in the spinal cord or peripheral nerves; (2) the fact that the muscular changes (atrophy and hypertrophy) do not, as a rule, seem to correspond in their distribution to the areas of distribu-

¹ *Clinical Lectures by German Authors.* Third Series (Sydenham Society's Translation), page 261.

tion of the spinal segments, nerve roots or peripheral nerves; and (3) the interesting fact pointed out by Babinski and Ohanoff (quoted by Grasset and Rauzier, page 643) that the muscular atrophy corresponds in its distribution to the 'embryonic areas' of the muscles.

Quite recently Von Babes has described certain alterations in the nerve terminations in the muscles in pseudo-hypertrophic paralysis and juvenile muscular atrophy, viz., (1) a defective development of the nerve-ending and of the fibre which gives rise to it; and (2) a degeneration (dissolution) of the already formed end-plate.¹ It will be extremely interesting to see whether these very suggestive observations are confirmed by independent investigation.

In some cases of pseudo-hypertrophic paralysis (and perhaps in the other varieties of myopathic muscular atrophy) there seems to be a tendency to a defect in development of the spinal cord. Now, if this is a fact (and I will refer to it more in detail when I come to describe pseudo-hypertrophic paralysis), it is of great importance, and suggests that the pathology of the so-called

¹ The following abstract of Von Babes' paper was published in the *Epitome of the British Medical Journal*, Oct. 6, 1894:—

THE NERVES IN PSEUDO-HYPERTROPHIC PARALYSIS.

Von Babes (*La Roumaine Médicale*, August, 1894) describes a new appearance (pathological) of the nerve terminations in this disease. Peripheral nerve terminations normally end in free blunt points or arborisations, and the recent discoveries with the Golgi method had served to emphasise this distinction. The author, in the course of examination of nerve endings in muscle and connective tissue, came across appearances which seem to differ essentially from the idea that nerve filaments necessarily have terminations in the strictest sense. In two cases—one of so-called primary juvenile muscular atrophy, and the other of infantile pseudo-hypertrophic paralysis—the following were found. One specimen of the pseudo-hypertrophic gastrocnemius with its connective tissue (fascia) was studied with Ranvier-Löwitt's gold chloride method, which stains the neuromuscular end plates. A remarkable change was observed; the motor fibre going to the muscle was in the first place remarkably thinned and attenuated, though preserving its myelin sheath, the latter being poor in cells and therefore presumably not undergoing degeneration. Its termination, where it abutted against the muscle fibre, was by a small conical enlargement, pale, and either simple or giving rise to a few very short and extremely fine filaments, which radiated from it star-wise. The terminal plate was thus of small size (atrophied or non-developed) and much simplified. [This appearance is indeed like those described by Ramon y Cajal in the growing points of the motor fibres of the spinal cord of the chick—'points d'accroissement'—in which case it would serve to show that the muscular nerve ending is yet in a very rudimentary condition

myopathic forms of progressive muscular atrophy should, as Erb has suggested, be founded on a much broader and more extensive basis than the purely myopathic origin of the disease would suppose.

The chief points of pathological and clinical difference between the myelopathic and the myopathic forms of progressive muscular atrophy.

Pathological differences. The essential pathological point of difference or distinction between the myelopathic form of progressive muscular atrophy (represented by the Aran-Duchenne type of the disease) and the myopathic forms (such as pseudo-hypertrophic paralysis, the juvenile form of progressive muscular atrophy of Erb, the facio-scapulo-humeral type of Landouzy and Déjerine) is that, in the former, the muscular atrophy is due to a lesion of nerve cells in the anterior horn of the spinal cord; whereas, in the latter, there is no discoverable lesion, either in the spinal cord or peripheral nerves. In the present state of our knowledge, then, the myopathic atrophy seems to depend upon a lesion which has its starting point in the muscles themselves.

in the disease.] Other specimens showed not only the atrophied (diminished) condition of the end plate, but the presence of small fusiform cells about this region, such as had been shown to be present in the experimental degeneration (Wallerian) induced in the nerve terminations by sections of the nerve trunk. This would seem to indicate that a veritable process of degeneration (dissolution) was going on. In two specimens (prepared by the isolation and teasing method) a single thick medullated fibre (4μ diameter) has been seen to divide, the two branches sweeping round in semicircular fashion to reunite into a distinct ring—a condition never observed before. From each segment of this ring or loop a fine medullated branch was given off, which, after winding spirally round the parent branch for some distance, left it to end freely; one of these shortly after ending in contact with a muscle fibre, the myelin sheath stopping short just before. The other fibre passed into the connective tissue, ending freely in an oval termination, within which the fibres formed a knot or swelling, and a number of sinuous twists without losing its continuity. This, the author thinks, is comparable to a sensory termination of the nerve; in any case, it is not motor. This double termination of a single medullated fibre, namely, by a motor and a sensory ending in the muscular tissue, the author is inclined to look upon as possibly normal, and which had before been overlooked owing to the extent and size of the nerve loop which gives rise to it. If confirmed, his conclusions would seem to show that the pathological conditions underlying the nerve ends in juvenile muscular atrophy and pseudo-hypertrophy are of the non-developmental type, that is (1) a defective development of the motor ending and of the fibre which gives rise to it, and (2) a degeneration (dissolution) of the already formed end plate, the two appearances being apparently distinct; while, on the other hand, the subdivision of a single muscular nerve into a sensory and a motor filament lands us in a new region of nerve function altogether.

Another point of pathological difference between the two forms seems to be this, that in the myelopathic form the affected muscles and muscular fibres simply waste; but in the myopathic forms the affected muscles almost invariably (probably always as a matter of fact at some period or other of their course) contain enlarged or hypertrophied muscular fibres. Whether this distinction is absolute or not is perhaps as yet uncertain; probably it is not absolute, but so far as our present knowledge enables us to judge, it is a point of great importance. And here I may say that if hypertrophied muscular fibres are occasionally met with in cases in which the nerve cells in the anterior horn of the spinal cord are undoubtedly affected, it lends some support to the view that the myopathic muscular atrophies are in reality myelopathic (see below). This does not of course necessarily follow. It is possible that in some of these cases the lesion is a mixed one. What I mean to say is this, that in some cases of progressive muscular atrophy it is perhaps possible that the myelopathic and myopathic forms of atrophy may be combined. Further, it has been suggested, though I cannot agree with this supposition, that the changes in the nerve cells are the result of the changes in the muscle, in other words, that the myopathic atrophy is the first event, and the atrophy and degeneration in the nerve cells of the anterior cornua a secondary result of that atrophy. There are, in short, various ways of looking at the facts. In the present state of our knowledge the question must, I think, be left an open one.

According to Erb, the histological changes in the muscles which are characteristic of the myopathic forms of progressive muscular atrophy are as follows:—

‘First of all there is very considerable hypertrophy of the fibres, to as much as three or four times the normal.¹ Then there

¹ ‘I know very well that, according to the observations of Auerbach and of Oppenheim and Siemerling, the existence of this hypertrophy *intra vitam*, at least in the fragments of muscle excised from the living subject, has been called in question. But as it is present in exactly the same way in muscles taken from the dead body, and as we can decidedly infer from the marked proliferation of nuclei and division of the fibres that there is some process of overgrowth going on in the muscle, I should like in the meantime to hold by the real existence of this hypertrophy, though its exact degree may possibly be modified and conditioned by the method of preparation. Knoll’s work has thrown some direct light on this point, and leads to the same conclusion.’

are all possible degrees of atrophy; rounding of the fibres till they are circular in form; increase of the nuclei both at the edge and in the interior; the formation of slits and the division of the fibres into two or more finger-like processes; vacuolation, sometimes only here and there, sometimes to a greater extent; faint transverse and pronounced longitudinal fibrils. On the other hand, there is no fatty or wax-like degeneration of the fibres.

‘The changes in the connective tissue are increase and overgrowth, slight to begin with, more abundant later on. There are firm broad strands lying between the muscle-fibres, with abundant nuclei and thickened multinucleated vessel-walls. Besides these changes there is sooner or later more or less of a deposit of fat, which may go on to distinct lipomatosis.’¹ (See fig. 58).

Clinical differences. The chief points of clinical difference and distinction between the myelopathic and myopathic atrophies are not perhaps quite so sharply and accurately defined as these pathological observations would seem to show; for according to some observers fibrillary twitchings and the reaction of degeneration, which Erb emphatically maintains are conclusive of the myelopathic form, are occasionally, though quite exceptionally, also present in the myopathic form. If subsequent observation should show that these statements are well founded—and I can speak positively as to the presence of fibrillary tremors, for in two cases of myopathic atrophy which have come under my observation the fibrillary tremors were extremely well marked—the fact may perhaps be explained, by supposing, either that (in these exceptional cases) there is a disordered or diseased condition of the nerve cells of the spinal cord, or that the lesion is a mixed one.

Speaking generally, the points of clinical distinction between the myelopathic and myopathic forms of progressive muscular atrophy are as follows:—

1. Fibrillary twitchings, which are such a characteristic and conspicuous feature in the great majority of cases of myelopathic atrophy (the Aran-Duchenne type and amyotrophic lateral sclerosis) are very rarely observed, in a characteristic and conspicuous form, in the myopathic forms.

¹ *Clinical Lectures by German Authors.* Third Series (Sydenham Society's Translation), page 258.

2. The reaction of degeneration, which in its imperfectly developed form is usually a characteristic feature of the advanced stages of the Aran-Duchenne type of progressive

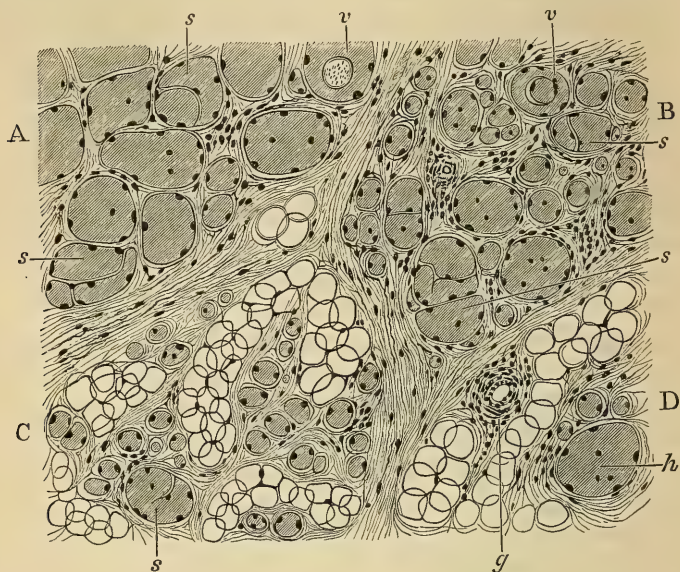


FIG. 58.—*Transverse section of muscle in progressive muscular dystrophy, made by combining four different sections from as many different cases.*—(After Erb).

A. From the hypertrophied deltoid of a typical juvenile case ; showing almost everywhere very much enlarged fibres with increase in the nuclei, slitting (*s*), and vacuolation (*v*) ; slight increase in connective tissue with abundant nuclei.

B. From the atrophied biceps of an indeterminate case (juvenile? pseudo-hypertrophic?) showing muscle-fibres of very different sizes, with great proliferation of the nuclei, abundant slitting (*s*), and here and there vacuolation (*v*) ; connective tissue very much overgrown and with numerous nuclei ; no fat deposit.

C. From the enlarged serratus magnus of a pseudo-hypertrophic case (our third case). Muscle-fibres mostly atrophied, here and there enlarged and slit (*s*) ; abundant firm connective tissue ; large deposits of fat cells.

D. From the very atrophied biceps of a juvenile case, showing little else than adipose tissue lying among stout bands of connective tissue, the vessels of the latter very much thickened (*g*) ; small insulated patches of muscle-fibre in some places still very much enlarged (*h*) with numerous nuclei.

muscular atrophy, is very rarely met with in the myopathic forms.

3. The small muscles of the hand, fingers and thumb, which are (? always) atrophied in the Aran-Duchenne type, and which,

in the great majority of cases, are the muscles which are first affected in that, the myelopathic, form of progressive muscular atrophy, are rarely affected, until, at all events, the later stages of the myopathic forms, though this may undoubtedly occur. It must, however, be remembered that in the later stages of the myopathic forms of muscular atrophy all the muscles, or practically all, may be atrophied.

4. True or false hypertrophy of the muscles is never met with in cases of myelopathic atrophy, but is very frequently developed in the myopathic forms of the disease. In pseudo-hypertrophic paralysis, the pseudo-hypertrophy of the muscles is one of the most striking clinical characteristics; indeed, from a diagnostic point of view, the pseudo-hypertrophy of the calf muscles is the most characteristic feature. In short, in the myelopathic (Aran-Duchenne) type of progressive muscular atrophy, the distal muscles of the limbs are usually first affected; while, in the myopathic forms of the disease, the proximal muscles of the limbs are usually first involved. In some cases of myopathic muscular atrophy the muscles of the back are first or very early involved. The frequency of this mode of development in the pseudo-hypertrophic type has I think in all probability been underestimated.

5. The myelopathic form of progressive muscular atrophy is very rarely hereditary; whereas, the myopathic forms are very strongly hereditary. Cases of the Aran-Duchenne type are almost always solitary, in other words, two or more cases of this disease are very rarely met with in the same family (brothers and sisters), or in the same generation (cousins); but it is quite common, in fact (so far as our present knowledge enables us to judge) the rule for more than one case of myopathic atrophy to be found in the same family or in collaterals of the same generation. I may, however, take this opportunity of stating that solitary cases of the myopathic forms of progressive muscular atrophy are probably more common than is usually supposed. As Erb has pointed out, the disease must have a beginning some time; and isolated cases are by no means uncommon. Nevertheless, the hereditary and family tendency is a very striking point of difference between the myopathic and the myelopathic forms.

6. The age at which the disease develops in the two cases is

different. The myelopathic form is essentially a disease of the adult and of the degenerative period of life; while the myopathic forms are usually developed during childhood, youth and early adult life, though in exceptional cases the disease may be developed later.

7. The order of development of the muscular atrophy is different in the two cases. In the great majority of cases of myelopathic atrophy (the Aran-Duchenne type), the small muscles of the hand are first affected; the disease very rarely commences in the lower extremities or in the muscles of the back, and the facial muscles are seldom if ever involved. Whereas, in the myopathic forms of atrophy, the small muscles of the hand almost always escape until the later stages of the disease; the atrophy often commences in the muscles of the lower extremities or sometimes in the muscles of the back (in the pseudo-hypertrophic form of the disease these are the usual starting-points), and occasionally in the muscles of the face.

8. In the myopathic forms of muscular atrophy, the tendon reflexes are diminished or abolished, never increased; whereas in the Aran-Duchenne type of the disease, the knee-jerks are often exaggerated and ankle-clonus is not unfrequently present.

9. In the Aran-Duchenne type of progressive muscular atrophy, 'bulbar symptoms' are sometimes developed, for the lesion may extend to the medulla oblongata; but, in the myopathic forms of muscular atrophy, bulbar symptoms do not occur.

The characteristic features of the myopathic forms of progressive muscular atrophy.—To sum up, the chief characteristics of the myopathic form of progressive muscular atrophy are as follows :—

1. The slow and gradual development of muscular atrophy and muscular weakness proportionate to that atrophy.

2. The progressive character of the muscular atrophy.

3. The development, though this is not invariable or at all events not always observable, of muscular enlargement, which in some cases appears to be a true and in other cases a false or pseudo-hypertrophy.

4. The absence of fibrillary twitchings.

5. The absence of the reaction of degeneration.

6. The strong hereditary tendency which the disease presents.

7. The strong tendency which the disease has to affect several members of the same family.

8. The fact that in the great majority of cases the disease develops during childhood, youth and early adult life.

9. The absence of any affection of the sphincters of the bladder and rectum.

10. The fact that if the deep reflexes are altered, they are always diminished or abolished, never exaggerated.

11. The entire absence of sensory symptoms.

12. The absence of any lesion which can be demonstrated by modern methods of investigation in the spinal cord, peripheral nerves, or (?) motor nerve-endings.

13. The presence in the affected muscles of enlarged (hypertrophied) muscular fibres.

As I have already pointed out, there is considerable difference of opinion amongst different observers as to the constancy of some of these characteristics. Some authorities, for example, state—and with them I agree—that in some exceptional cases of myopathic atrophy, fibrillary tremors occur: others say that the reaction of degeneration is sometimes developed in some of the affected muscles. Further, it is doubtful whether the presence of enlarged (hypertrophied) muscular fibres is absolutely pathognomonic.

The muscular atrophy and muscular enlargement (either true or false) which constitute the essential clinical feature of the various forms of myopathic muscular atrophy, may develop in a different order and be distributed in a different manner in individual cases of the disease. Several forms or types (based upon the order in which the muscular atrophy is developed and the manner in which the muscular atrophy and pseudo-hypertrophy are distributed) have been described. Until quite recently these different varieties or types were considered to be different diseases; but recent observations seem to show that many of them, at all events, are mere modifications or varieties of one fundamental or primary disease—the myopathic form of progressive muscular atrophy or progressive muscular dystrophy, as it has been termed.

The facts which Erb has advanced in favour of the view that

the different types of myopathic muscular atrophy are modifications of one and the same disease are in my judgment conclusive. He says:—

‘They all agree in the slow and insidious development of the disease, with very frequently hereditary or family influences at work; in the general wasting of many of the muscles combined with increase in the size of others; very notably in the localisation of the affection, in that almost invariably they are exactly the same muscles in the different cases that atrophy on the one hand, and exactly the same that become hypertrophied on the other; in the way in which this localising takes place in the trunk, shoulder girdle, and loins, and in the proximal ends of the limbs, the distal ends remaining free for a long time; and in the peculiar changes which are then brought about in the form and movements of the body, the attitude, and gait. They agree further in the condition of the muscles, as ascertained by percussion and palpation, in the effects of electrical stimulation, particularly in the absence of the reaction of degeneration, in the fact that there are no fibrillar twitchings, and in the gradual disappearance of tendon reflexes. Finally, in all of them the various forms of skin sensibility, the special senses, the muscular sense are absolutely normal, the sphincters are unaffected; the brain and organs of special sense, and all the internal organs, are quite normal.

‘Thus in all the forms the outline of the features is the same, and yet there are certain differences which cannot be overlooked. These are as follows: Some of the cases appear to be independent of hereditary influences; sometimes the process begins in earliest childhood, sometimes in youth or at puberty, sometimes even later. In some cases it makes its appearance first in the loins and lower extremities; in others in the shoulders and upper extremities; at times even in the face. The rules of localisation are often set aside, the muscles of the forearm and small muscles of the hand being sometimes attacked, and muscles which, as a rule, are affected, being sometimes exempt. The degree of muscular hypertrophy may vary extremely, at times being quite unnoticeable, or limited to a few muscles, at times extended over a wide area, and present in a very marked degree. Further, this hypertrophy appears in some of the cases to be a true hypertrophy, depending on actual increase in volume of the muscle substance, and in others to be false, caused by a deposit of fat or lipomatosis. Lastly, the rate at which the disease advances, and the way in which, towards the close, all the muscles of the body may be affected, are very different in the individual cases.

‘The longer I have occupied myself with the question, and the wider my experience of these forms has grown, the more has the conviction forced itself upon me that they all present one and the same disease. I am satisfied that, while in subordinate features such as the time and rate of development, the initial localisation, quantitative differences in the individual symptoms, particularly as regards the amount of hypertrophy, they may differ from one another, yet in all essential points they thoroughly agree. The proof of this clinical unity of those forms must, in the first instance, of course, be deduced from clinical material, but, from the number of my own cases and from a record on the part of others which has gradually grown to be very large, we have no difficulty about that.

‘First of all we must establish the fact that the separate forms agree with one another in the following particulars:—The development and the localisation of both the atrophy and the hypertrophy in the muscles; the condition of the latter, as ascertained by inspection and palpation, by their mechanical and by their electrical reaction; and, lastly, the absence of all other symptoms.

‘But still more convincing evidence is given by the cases, and they are not so very rare, which may be looked upon as transitional varieties between those individual types—cases of one form in which you meet with certain features which you have learned to consider as properly belonging only to another form. For example, there have been lately observed several cases, both of the juvenile and of the pseudo-hypertrophic form, in which the muscles of the face have become involved (as in the second case which I showed you); or you may see the infantile form, beginning with pronounced facial atrophy, developing, as regards the rest of the body, at one time the juvenile type, at another time the pseudo-hypertrophic type; or you may observe the lower half of the body take on the distinct pseudo-hypertrophic type, while the upper half is an example of simple atrophy—the juvenile type; or you may have the exact appearances of pseudo-hypertrophic paralysis, coming on in adult life, i.e., as a juvenile form.

‘Further, we not infrequently see the different forms passing into one another in the course of their development. A case that began as pseudo-hypertrophy takes afterwards the juvenile or infantile form; a hereditary case turns into a pseudo-hypertrophic or juvenile case, and so on. We frequently come across cases, also, which I should like to call indeterminate or, better, indeterminable forms, cases in which there may be doubt as to which type they belong to. We have seen an example of this in our fourth case, and to a certain extent in the two sisters

(cases 5 and 6). In a case of this kind one man sees one type, another another; or perhaps the case has been taken for a certain type at one time and two years later the physician finds himself inclined or obliged to call it an example of another. And yet all these cases most certainly belong to the same nosological group.

‘Different types occur in the same family, and this fact, it appears to me, speaks with great force for the view we are upholding. For in that case they occur in circumstances where there is no room for doubt as to the unity of the disease. For example, the infantile form may appear among children whose father has the juvenile form (observed by Duchenne, Landouzy-Déjerine, Troisier-Guinon and others), or different types may occur in a family in which the hereditary form has already gained a footing. These are, I think, very convincing proofs.

‘I believe that the facts I have laid before you in the present state of our knowledge, and disregarding just now the proof from pathological anatomy, to which I shall return later, are sufficient to allow you to recognise in all the different types one nosological species. It is fitting that we should have for this a short distinctive name, and I proposed as such *Dystrophia muscularis progressiva*. I still think that it is the best, and that it involves fewer assumptions than any of those that have been proposed by others.’^{1 2}

Let us now consider the special features of these different varieties or types in more detail.

Classification and Varieties.—And first let me say a word or two with regard to classification.

The more important types or varieties of myopathic atrophy which have been described are as follows:—

1. *Pseudo-hypertrophic paralysis*.

In this form, the muscular weakness and atrophy commence in the muscles of the legs or back, and the calf muscles are enlarged from pseudo-hypertrophy.

2. *The ‘juvenile form’ of progressive muscular atrophy of Erb.*

¹ ‘The name chosen by Fr. Schultze, “Progressive muscular wasting associated with hypertrophy,” is too cumbrous, and scarcely suitable for others than Germans. The “Myopathie atrophique progressive” of Landouzy and Déjerine takes no account of the hypertrophy that is generally present. Charcot’s “Myopathie progressive primitive” would suit best of all if it were only certain that the myopathy is primary.’

² *Clinical Lectures by German Authors* (Sydenham Society’s Translation), page 247.

In this form, the atrophy usually commences in the muscles of the shoulder girdle and upper arm, and the deltoids (amongst other muscles) are enlarged.

From the following passage it would appear that Erb intended to include under the term 'the juvenile form of progressive muscular atrophy' all cases of progressive muscular atrophy of myopathic origin. He says:—

'At that time (1883), grounding on numerous cases of my own, I made an attempt to show that this apparent unity contained at least two clinically and probably anatomically different forms of disease, and that these were clearly distinguishable throughout in symptoms, development, localisation, and actual condition of the tissues. By the side of the well-defined spinal form, which depends on lesion of the anterior horns of grey matter, I placed another, which I called 'juvenile' muscular atrophy, and I sought then to show in detail that the pseudo-hypertrophic paralysis and the hereditary form of Leyden both belong to the latter type.'¹

3. *The 'facio-scapulo-humeral type' of progressive muscular atrophy* of Landouzy and Déjerine, which had been previously described by Duchenne as the *infantile form of muscular atrophy*.

In this form, the atrophy commences in the muscles of the face and may subsequently involve the muscles of the shoulder girdle, upper arm, back, etc.

4. *The hereditary form of progressive muscular atrophy* of Leyden.

In this form the muscular weakness and atrophy commence in the muscles of the back or lower extremities; the calf muscles are often enlarged.

5. *The (purely)² atrophic form of myopathic muscular atrophy*.

In this form, the muscular weakness and atrophy commence in

¹ *Clinical Lectures by German Authors*, Third Series (Sydenham Society's Translation), page 235.

² I refer to the clinical characters. It is probable that in some of the cases in which there is no obvious hypertrophy when the patient comes under the notice of the physician, some muscular enlargement had previously been present; and that in other cases, there is some relative enlargement; the calves, for example, though not actually enlarged, may be relatively large in comparison with the thighs or upper limbs.

the back or lower extremities; the cases only differ from Leyden's type in this particular that the calf muscles are not enlarged.

Identity of these different forms.—Almost all authorities are now agreed that Erb's juvenile form and the facio-scapulo-humeral type of Landouzy and Déjerine are mere varieties of the same condition.

The hereditary form of Leyden and the cases which I am in the habit of terming the purely atrophic form are undoubtedly mere varieties of pseudo-hypertrophic paralysis.

Further, it seems to me highly probable, as Erb has suggested, that all of these forms are mere modifications or varieties of the same disease. The differences which seem at first sight to exist between pseudo-hypertrophic paralysis, on the one hand, and the juvenile form of myopathic atrophy and the facio-scapulo-humeral type of Landouzy and Déjerine, on the other, are probably not real points of distinction.

6. *The diffused form of progressive muscular atrophy of infancy and early childhood.*—As I shall afterwards point out, this appears to be a distinct clinical type. It is doubtful whether it is a myopathic or a myelopathic form. Personally, I am disposed to agree with the latter view, and to think that the cases which are usually grouped under this head are probably identical with those described by Duchenne under the term *paralyisie générale spinale antérieure subaiguë* (see page 111).

7. *The peroneal type of progressive muscular atrophy.*—This form is undoubtedly distinct from the others enumerated above. Its exact pathology is still undetermined. Some writers have supposed that it is a myopathic, others a neuropathic, and others again a myelopathic, atrophy. As I shall presently tell you, in some of its clinical features it seems to bear a close resemblance to the Aran-Duchenne type of progressive muscular atrophy—a fact which, so far as I know, was first insisted upon by Sachs.

The classification, then, of the forms of progressive muscular atrophy enumerated above which, in the present position of our knowledge, seems to me the most satisfactory, is as follows:—

1. *Myopathic atrophies (Progressive muscular dystrophies).*

Varieties:—(a) Pseudo-hypertrophic paralysis.

(b) Juvenile form of progressive muscular atrophy of Erb.

- (c) Facio-scapulo-humeral type of Landouzy and Déjerine.
- (d) Hereditary form of progressive muscular atrophy of Leyden.
- (e) The (purely) atrophic form of progressive myopathic atrophy.

2. *The diffuse form of progressive muscular atrophy of infancy and early childhood.* (Probably myelopathic.)

3. *The peroneal type of progressive muscular atrophy* (Probably neuropathic or myelopathic.)

Let me now describe in more detail the clinical features of these different varieties, and particularly of pseudo-hypertrophic paralysis, for it is by far the most common form.

LECTURE XIV

PSEUDO-HYPERTROPHIC PARALYSIS

PSEUDO-HYPERTROPHIC paralysis, to which the terms *Atrophia musculorum lipomatosa*, *Lipomatous myo-atrophy*, *Pseudo-hypertrophy of muscles*, and several other synonyms have been applied, is a very interesting and somewhat rare disease.

It is essentially characterised by slowly developing muscular weakness and muscular atrophy, and by the enlargement of some of the muscles which are undergoing the atrophic process. The increased size of the muscles is not a true hypertrophy, but is due to the production in the muscles of an excess of fat and fibrous tissue. Hence the term *pseudo-hypertrophic paralysis*.¹

Morbid Anatomy.—In its slow development, progressive course, and the marked muscular weakness and muscular atrophy by which it is characterised, pseudo-hypertrophic paralysis resembles the Aran-Duchenne type of progressive muscular atrophy; and, as I have already remarked, it was at one time supposed that, like the spinal form of progressive muscular atrophy, the disease was due to a lesion of the anterior cornual region of the spinal cord. I need not again discuss this question in detail. Suffice it to say that it is now generally believed that the muscular wasting is myopathic; that is to say, that it is due to an affection of the muscular tissue itself, and that it is not the result of a lesion of the spinal cord or peripheral nerves.

¹ Recent observations would seem to show that this statement cannot be absolutely applied to all cases. Erb, for example, states with regard to one of his cases:—‘According to former usage this case also would have been termed pseudo-hypertrophic paralysis without much ado, although as you have satisfied yourselves, the muscles that are enlarged and are acting well by no means give the impression of pseudo-hypertrophy or lipomatosis.’

Pseudo-hypertrophic paralysis, though not a very rare, is by no means a very common disease; and the opportunities for investigating the pathology after death seldom occur. The number of cases which have been examined after death is surprisingly small—out of proportion small to the frequency with which the disease occurs. In proof of the statement, that the disease is not very rare, I may mention that at the present

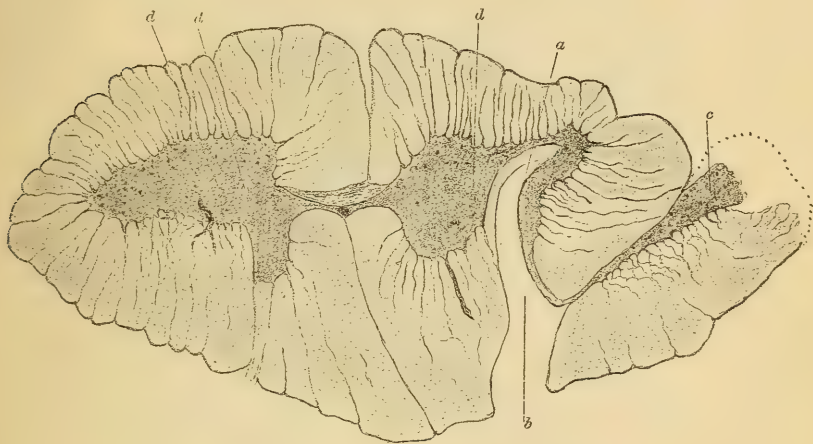


FIG. 59.—*Transverse section through the middle of the cervical enlargement in a case of pseudo-hypertrophic paralysis. Stained with carmine, mounted in dammar, very low power.*

The grey matter of the right lateral half is split by a band of white matter (*a*) which passes into it from the posterior column, and by a deep fissure (*b*); *c*, detached portion of grey matter running out to the surface of the lateral column; *d, d, d*, fissures or tears in the grey matter.

time I have eleven cases under observation—two in Edinburgh and nine in other parts of the country. The paucity of autopsies is no doubt explained by the facts that the patients very rarely die in hospital, and that post mortems involving the examination of the spinal cord are seldom made in private practice.

In the great majority of the (undoubted) cases of pseudo-hypertrophic paralysis which have been examined after death, the spinal cord was perfectly normal; and in the comparatively few cases in which the peripheral nerves seem to have been

carefully examined, there was no lesion in this part of the nervous apparatus.¹

In the few cases of pseudo-hypertrophic paralysis in which pathological changes were present in the spinal cord, the alterations were probably associated or accidental lesions.



FIG. 60.—Transverse section through the lumbar enlargement of the spinal cord in a case of pseudo-hypertrophic paralysis (the case represented in figs. 78 and 79), showing a fissure, to which the letter *a* points, running into the posterior horn of grey matter.

The tissue of the posterior horn is disintegrated. The membranes are detached from the surface of the posterior columns. Numerous nerve cells are seen in the anterior cornua.

In one case which I examined some years ago, the grey matter was curiously malformed in the cervical region (see fig. 59).

In another case which has proved fatal since this lecture was delivered (the case represented in figs. 78 and 79), the spinal cord is also misshaped. A fissure dips down into the posterior horn of grey matter on the left side of the lumbar enlargement (see figs. 60 and 61). I first became aware of the presence of this fissure when I came to cut up the partially hardened cord into suitable pieces for microscopic examination. Not

¹ On this point see page 204.

suspecting any naked eye lesion, I had not carefully examined the exterior of the cord in the fresh state. The fissure was quite apparent to the naked eye. The intermediate grey matter is disintegrated and the distribution of the nerve cells in the posterior part of the cord (posterior column and posterior part of

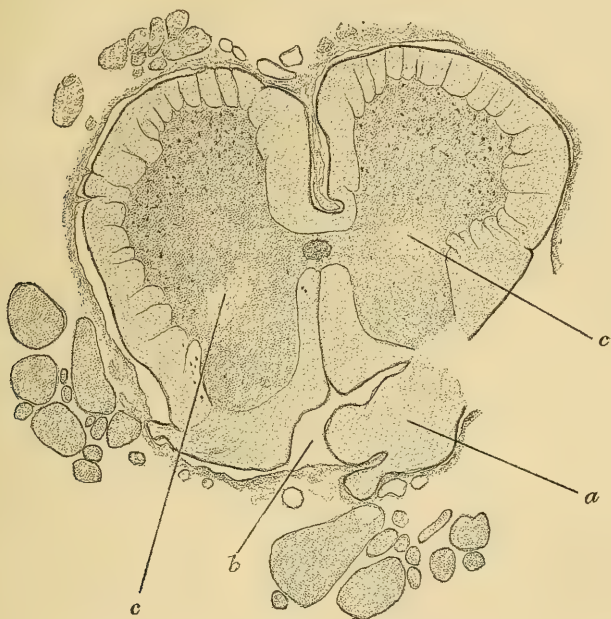


FIG. 61.—*Transverse section of the same cord (fig. 60) at a lower level.*

The posterior column is peculiarly shaped ; part of the posterior horn of grey matter and a knob-shaped mass of the posterior column are separated from the rest of the cord by an artificial fissure. The intermediate grey matter on the right side is disintegrated. Two nerve cells are seen in the left posterior column and several in the left lateral column just outside the posterior horn of grey matter.

Note.—In the process of mounting this section has been reversed ; the right half of the drawing is really the left half of the cord.

the lateral column) is, so far as my experience enables me to judge, peculiar. Two sections of the cord which Dr. Muir has kindly prepared for me are represented in figs. 60 and 61. I feel great hesitation in expressing a positive opinion as to the significance of the changes which are present in these sections. They can only be the result either (*a*) of artificial damage to the cord, say by the point of the bone pliers in removing it from the

body; (*b*) of congenital malformation; or (*c*) of disease. The fissure (whether originally due to artificial damage or to disease) may no doubt have been exaggerated and enlarged by the process of hardening.

That some of the changes are artificial I have little doubt, but that they are altogether due to this cause is by no means clear. I fail to see, for example, how the condition represented in Fig. 61 could be altogether the result of artificial damage. Dr. Muir takes a different view. After very careful examination, he concludes that there is no structural alteration which must necessarily be looked upon as *ante mortem*.¹

Professor Sherrington, who has also kindly examined the sections, admits the great difficulty there is in forming an opinion as to the significance of the alterations, and states:—‘I do not think the cavity in your preparation explicable as an *ante-fact*.’²

¹ In the report which Dr. Muir has kindly furnished me with, he says:—‘What is the nature of the fissure? In favour of part at least being natural is the regularity of the margins and the condensation, and so much is this the case at places that it is difficult to conceive of its being produced by artificial means. But, on the other hand, so far as I can find after careful examination of the edges, there is no structural alteration which must necessarily be looked upon as *ante mortem*. The surface of the cord also shows signs of injury posteriorly, in the stripping of the pia, displacement of the nerve roots, etc. My impression at first was that there had probably been an elongated area of softening in the posterior cornu which had been artificially extended to the surface, but this supposition would not explain the position of the left posterior column at the higher level described, i.e. before the fissure reaches the surface. At this level the left posterior column is much smaller than the right. It must either have been so naturally—which is not the case at higher and lower levels in the cord—or a portion must have been shaved off, when the pia was removed. So far as I can see, it is impossible to come to an absolute conclusion, but I do not think there is sufficient evidence from the sections alone to enable one to describe the changes as being undoubtedly *ante mortem*.’

² Professor Sherrington’s full report is as follows:—

‘The specimen was certainly well hardened, an important point in judging of the character of the fissure and cavity. I think some of these fissures offer the hardest problems in morbid histology.

‘The appearance of a condensed edge can be sometimes given and closely resemble a sclerosed edge even in a purely artificial cavity. Some years back I made a number of artificial cavities in portions of spinal cord purposely by bending the pieces of cord at a not very sharp angle and tying them in that position and then hardening in Müller’s fluid. The cavities obtained nearly always affected the posterior column and always ran into the grey matter opening there into a larger space. The sections through such *ante-facts* do not, however, agree in character with the appearance in your specimen. I do not think the cavity in your preparation explicable as an *ante-fact*.

Further, somewhat similar alterations have been described by independent observers.

In a case reported by Drummond, the grey matter of the anterior horn, at its junction with the posterior cornu and in its lateral part (*the same situation affected in my cases*) was softened and disintegrated, so that a cavity was formed in the centre of the cord; this cavity, which had no proper cyst wall, was of large size in the lumbar enlargement, where it caused the cord to bulge out laterally, and extended through the dorsal and cervical regions; 'with a high power, minute disintegration could be traced through the lateral grey net-work of both sides, the degenerative appearances being most conspicuous around the blood-vessel'.

In another case (which was examined by Gowers and Lockthar Clarke) 'at the lower dorsal segment, there was an area of granular disintegration in the intermediate grey substance on each side, in front of the posterior vesicular tract (*the same portion of the grey matter which was affected in my two cases and in Drummond's case*). This part was unduly translucent for half a centimetre in vertical extent, and in the middle of this area the disintegration had produced an actual cavity, across which the fibres for the cerebellar tract ran unchanged.'¹

'By hardening in bichromate we sacrifice the grey matter—except the borders of the coarser ganglion cells in it—for the sake of the white column. The spongiosa is shrunken much more than the white matter, as can be seen by its sunken level at the ends of a block of cord hardening in Müller. Also the connective tissue shrinks (even the tough dura) much more than the white matter. If a short length of cord with dura slashed transversely over one half and left without transverse cuts over the other half, be placed in bichromate, an hour's time is enough to curve it strongly over toward the side of the unslashed dura.

'Cavities which are ante-facts are, I am convinced from what I saw at that time, due to unequal shrinkage of grey matter and connective tissue on the one hand and of white matter on the other, resulting in rending and cleavage. The cavity in your preparation gives me, on detailed examination, the impression that it has extended to the surface in the hardening. I am led to think there was no fissure which reached the cord in the fresh state, but that in the depth of the posterior horn, or posterior part of grey matter further forward, there was either an actual cavity or a region of altered, loosened, atrophic tissue which, under hardening and dehydration, shrank greatly, splitting as it did so the weakest part of the wall of tissue adjoining. I imagine this happened when in the bichromate and before the preparation was placed in spirit, because the tissue bounding the cavity and cleft bear marks of having been particularly under the action of bichromate.'

¹ Gowers' *Diseases of the Spinal Cord*, Second Edition, p. 516.

It is possible that the changes in the intermediate grey matter which were present in all of these cases were the result and not the cause of the disease. But in my first case, the malformation of the grey matter was clearly congenital. I do not, of course, suggest that this malformation was the cause of the lesions in the muscles; but I am strongly inclined to think that it was not merely accidental. It suggests that in pseudo-hypertrophic paralysis there is a widespread tendency to congenital or developmental defects in the neuro-motor apparatus—not merely in the muscles but in the spinal cord and perhaps (as Von Babes' observations to which I have already referred on p. 204 would seem to show) in the nerve-terminations in the muscles.

In my second case (represented in fig. 60) the multipolar nerve cells in the anterior horn were numerous and large, many of them were slightly fatty and their nuclei somewhat less distinct than normal.

So far as I know, there are no facts either clinical or pathological (except Von Babes' observations) to support the view that the morbid process in the muscles has its starting-point in the peripheral nerves. Von Babes' observations, if confirmed, would seem to show that the end-plates and fine terminations of the motor nerves in the muscles are imperfectly developed or degenerated. The facts that in cases of pseudo-hypertrophic paralysis the muscular co-ordination is so remarkably perfect and that there is very rarely, if ever, any trace of the 'reaction of degeneration' in the affected muscles are perhaps difficult to reconcile with this (Von Babes') view.

The very remarkable way in which patients affected with pseudo-hypertrophic paralysis can co-ordinate their movements and balance themselves in the erect position, even when their muscular power is very greatly enfeebled, seems conclusively to show that there is no interference with the muscular sense and with the ingoing impressions concerned in muscular co-ordination, which pass from the muscles to the spinal cord. We may confidently, I think, conclude that the sensory nerves of the muscles are entirely unaffected.

Further, as I shall afterwards point out, the sensory functions of the skin are absolutely uninterfered with. The defect, whatever it may be, is clearly confined to the motor or trophic-motor side of the nerve apparatus or to the muscles themselves.

The histological changes in the muscles.—The most striking pathological changes in the muscles after death are (1) an overgrowth of the fatty and interstitial fibrous tissues, and (2) an atrophy of the muscular fibres themselves. In advanced cases, the muscular tissue may, on naked eye examination, seem to have completely disappeared and the muscles may look like masses of fat.

On microscopic examination, the muscular fibres and bundles of muscular fibres are seen to be separated by large masses of fat cells and an excessive quantity of interstitial connective tissue. (See figs. 62 and 63.)

In the advanced stages of the disease, many of the muscular fibres are markedly atrophied. The atrophy seems in part to be a simple atrophy, in part a degenerative process. Many of the most minute (atrophied) fibres still retain their transverse striation, while others are degenerated, vacuolated, split up longitudinally or in process of transformation into fibrous tissue.

Some of the muscular fibres are enlarged, but this change (hypertrophy of the individual muscular fibres) is much less conspicuous in pseudo-hypertrophic paralysis than in some of the other forms of progressive muscular dystrophy.

These appearances are well seen in figs. 62 and 63. The connective tissue and blood vessels between the atrophied fibres are usually very rich in nuclei. So far as my observation enables me to judge, this change is especially marked in the earlier stages of the disease. In the later stages, the interstitial tissue between the wasted fibres consists chiefly of fat cells.

As I have already remarked, some authorities suppose that the atrophy of the muscular fibres is secondary to the interstitial changes; and, so far as I can judge, this is partly true, but I doubt if it expresses the whole truth. The weight of clinical and pathological evidence seems to me to be in favour of the identity of this (the pseudo-hypertrophic) and the other forms of myopathic atrophy; and it must I think be acknowledged that in the non-pseudo-hypertrophic forms of myopathic atrophy the wasting of the muscles is chiefly due to a degenerative process, and is not merely the result of a simple atrophy from pressure.

In connection with this point it must be remembered that too much importance should not be attached to the examination

of the muscles in old-standing cases of pseudo-hypertrophic paralysis. In many of the cases which have been examined, the disease has been in existence for many years and at the time of death the final stage of atrophy has been reached. For the purpose of demonstrating the mode of development of the

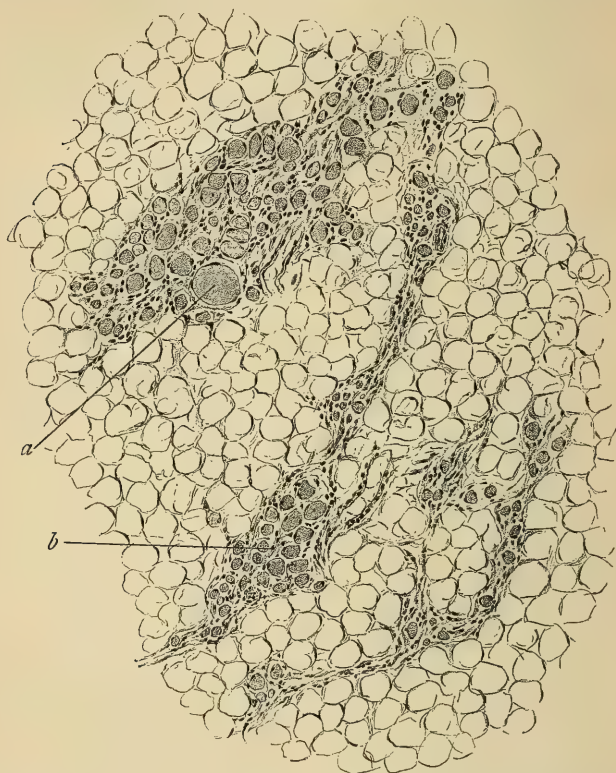


FIG. 62.—*Transverse section of the calf muscle in pseudo-hypertrophic paralysis (the case which is represented in fig. 78).*

The drawing was made from a section prepared by Dr Muir.

muscular changes, the condition of the muscles in the early stages of the disease is far more important than the condition in the last stages of the atrophy.

In this, as in other forms of myopathic atrophy, the enlargement (apparent hypertrophy) of certain muscles (the calf muscles and the spinati muscles, for example) is associated with atrophy

of other muscles. This combination of atrophy and apparent or pseudo hypertrophy is a characteristic feature of the disease. So far as one can judge from the condition of the muscles during life, the atrophy in some of the muscles is a simple atrophy. In many of the muscles, it is not, so far as one can see with the naked eye, preceded by any enlargement. But after death, the muscles which have apparently undergone this

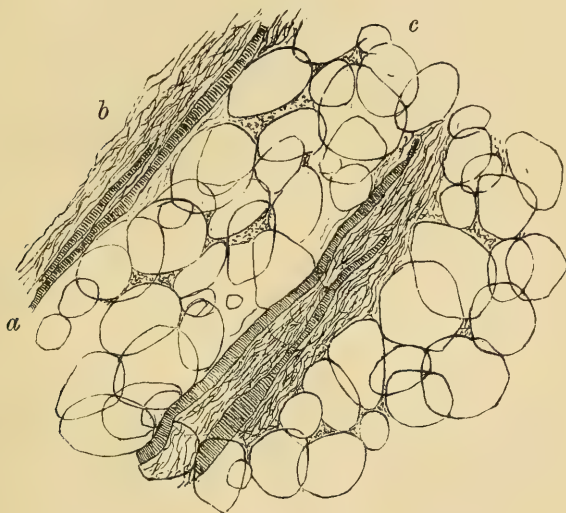


FIG. 63.—*Longitudinal section of muscle in pseudo-hypertrophic paralysis.*—
(After Gowers.)

a atrophied muscular fibre, with transverse striæ still remaining ; *b*, band of fibrous tissue ; *c*, fat cells.

simple atrophy are in many cases infiltrated with fat and similar, so far as their histological characters are concerned, to the muscles which were formerly enlarged.

These pathological changes are not entirely confined to the voluntary muscles ; for in one of my cases the muscular tissue of the heart was affected. (See fig. 64.)

In the advanced stages of old-standing cases (see fig. 78) the subcutaneous fat may entirely disappear just as it does in the advanced stages of progressive muscular atrophy. But the

development of the bones, in length at all events, does not seem to be interfered with. This is a point of some interest.



FIG. 64.—Section through the wall of the left ventricle of the heart in pseudo-hypertrophic paralysis, showing degenerated muscular fibres in the midst of fibrous tissue.

Etiology.—Pseudo-hypertrophic paralysis is essentially a disease of childhood, and is much more common in boys than in girls. The disease usually commences before the sixth or seventh year. In some cases, the symptoms are first noticed when the child begins to walk; in others, the onset does not occur till later childhood; in rare cases, the symptoms are first noticed at or about the time of puberty. In the, comparatively speaking, rare cases in which the disease appears to commence after the development of the body has been completed (at or after the age of 20) it is usually mild in type and has in all probability been slowly and insidiously progressing for some time before it was sufficiently marked to attract the attention of the patient. In girls, the disease is usually developed later than in boys; and in girls, the disease is usually less severe than in boys. In fact, speaking generally, we may say that the earlier the disease is developed, the more severe is its type and the more rapid its progress; and, *vice versâ*, the later it is developed, the less severe is its type and the less rapid its progress. There are of course exceptions to all of these statements.

Further, it seems probable that the later the disease is developed the more likely it is to assume the form which has been described by Erb as the juvenile type of the disease.

The disease usually begins insidiously without any apparent cause. In rare cases, the onset is preceded, or appears to be preceded, by an attack of measles, scarlet fever, or some other infantile complaint; but in many of the cases in which measles or other acute diseases appear to the parents to be the cause, the disease was in all probability already present and unrecognised. At the time of birth and during early childhood (*i.e.* up to the onset of the disease), the patients present no peculiarity; they are usually quite healthy up to the time at which the disease develops; but in other cases they are longer in walking than most healthy children.

Pseudo-hypertrophic paralysis is essentially a 'family disease'; it is quite common to find several members of a family, usually several brothers, less frequently the sisters as well as the brothers, affected. The disease is clearly due to something which is born with the individual—to something which is handed down from the parent to the children. The disease is rarely directly hereditary (*i.e.* the parents of children who manifest the disease are very rarely themselves affected); but the uncles, aunts, grand-uncles or grand-aunts, and (far more frequently) the male cousins may have been affected. In several of my own cases, the hereditary tendency could not be traced, and in at least three instances one member of a family only was affected. It is of course possible, indeed probable, that in some of these cases other members may ultimately become affected.

When successive cases occur in different generations, the disease is almost always passed on through the female line. Exactly the same thing is seen in Friedreich's ataxia. It necessarily happens that pseudo-hypertrophic paralysis can rarely be directly handed on from parent to child; for the males who are affected with the disease are usually helpless cripples by the time of puberty, and females who are affected are not likely to be chosen in marriage, although in them the disease is often only slight when they attain the marriageable age. But this statement only affords a partial explanation of the fact that the disease is almost exclusively handed down through the female line; for the male children of the healthy brothers of pseudo-

hypertrophic patients usually escape, while the male children of the healthy sisters are often affected.

It is doubtful whether a neuropathic inheritance has much to do with the production of the disease.

Some writers state that the disease is as common in the upper as in the lower ranks of society, but this is not my experience; with very few exceptions my cases have occurred in hospital patients.

Clinical History.—The essential clinical feature of pseudo-hypertrophic paralysis is a slowly-developing and gradually-increasing muscular weakness, with enlargement of some and atrophy of other muscles.

As I have already told you, the disease usually begins in childhood, in the great majority of cases before the seventh year. In some cases, the muscular defect (weakness) is noticed from the time that the child begins to walk. The first symptom to attract attention is usually clumsiness and insecurity in the gait; the child stumbles or falls without sufficient cause. In some cases the parents are astonished that the child is so weak on its legs; for, as they will sometimes tell you with pride, 'its calves are finely developed.' As the disease progresses, the gait becomes waddling in character.

Duchenne divided the disease into three stages. The *first* characterised by difficulty in standing and walking, and weakness of the muscles of the lower extremities and of the sacro-lumbar region; the *second* by muscular hypertrophy; and the *third* by increasing feebleness of the muscles both of the upper and lower extremities. But these stages are only roughly speaking and approximately accurate; for modern observations have abundantly shown that at the same time that some of the muscles are enlarged (apparently hypertrophied) others are atrophied. It is important to note that many of the muscles which are atrophied never present any previous stage of enlargement or pseudo-hypertrophy.

The first symptom is usually weakness in the muscles of the back and legs. As the disease progresses and the weakness becomes more marked, the back becomes curved, the attitude and gait become characteristic, and the child has difficulty in rising from the recumbent to the erect position.

The attitude.—In well marked cases (i.e. when the disease is

typical and fully developed), the patient usually stands with his feet wide apart (see figs. 65 and 66) so as to enlarge his base of



FIG. 65.



FIG. 66.

A typical case of pseudo-hypertrophic paralysis showing the attitude in the erect position.

The patient could stand and walk, but the slightest push, which upset his centre of gravity, was sufficient to throw him down.

The back is markedly curved and the chest thrown forwards.

The infraspinati muscles are enormously enlarged. The masseter muscles were also enlarged.

The patient was unable to raise himself completely from the recumbent to the erect position.

support as much as possible; the heels are in many cases drawn up in consequence of retraction of the calf muscles (see fig. 67); the back is strongly curved in the lower dorsal and lumbar regions (lumbar lordosis), the shoulders are drawn far back and the abdomen is prominent; in some cases (as Duchenne has pointed out) a line drawn vertically downwards from the middle of the shoulders falls behind the sacrum (see figs. 68 and 69). In many cases the chest is flattened in the antero-posterior direction.



FIG. 67.—An advanced stage of pseudo-hypertrophic paralysis, showing the position of the feet in the erect position of the body.

When the patient stands erect, or when he attempts to walk, the arms are usually extended by the sides and used to balance the body. The attitude is a very striking one, and it is remarkable how, even in the comparatively speaking advanced stages of the disease when the muscular debility is extremely great, the patient is able to maintain his balance in the upright position. As I have already remarked, there is no defect of co-ordination; in fact, the patient's power of balancing the body and adjusting his equilibrium under the very unfavourable circumstances in which he is placed in consequence of the muscular weakness is a remarkable feature of the disease. But although the patient can

stand and maintain his equilibrium so long as he is undisturbed, he is extremely insecure; the slightest push in any direction is sufficient to throw him over; a mere breath of wind is almost sufficient to knock him down; and if his centre of gravity is displaced he falls down with a thump, all of a heap as it were.

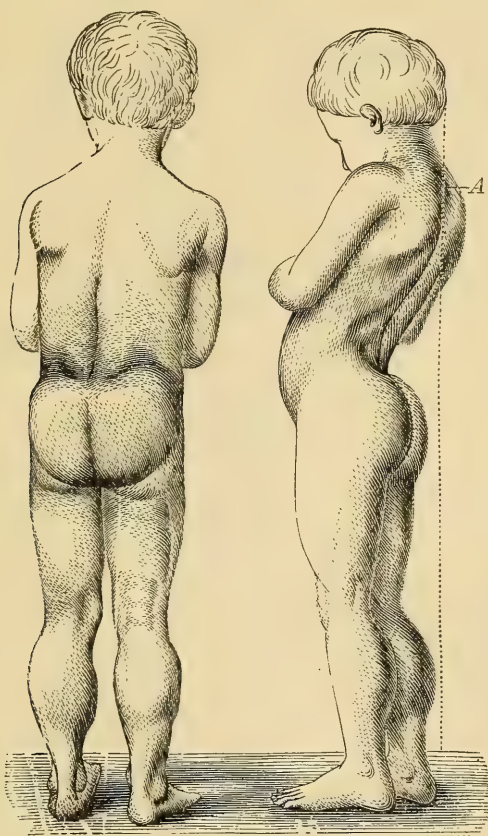


FIG. 68.

FIG. 69.

Pseudo-hypertrophic paralysis, showing enlargement of the calves and buttocks. The back is curved, and a line drawn downwards from the scapula falls behind the sacrum.—(After Duchenne.)

The antero-posterior curvature, which is such a striking feature when the patient stands erect, entirely disappears when he assumes the sitting position. Duchenne believed that this curvature was due to weakness of the spinal muscles; but some

authorities think that it is rather the result of weakness of the extensor muscles of the hip.

The gait.—In most cases the gait is, even in a comparatively early stage of the disease, very characteristic. The body oscillates or sways from side to side; the gait is waddling in character and in some cases high-stepping. There is great difficulty in flexing the thigh on the abdomen, in projecting the thigh forwards and in raising the toes from the ground; consequently even at a comparatively early stage of the disease (i.e. when the patient can walk on the flat) he is unable to go up-stairs. He has to drag himself up by the handrail. In many cases even in the early stages of the disease, the patient cannot raise himself on his toes and cannot jump. In the great majority of cases of pseudo-hypertrophic paralysis it will be found on inquiry that the patient has never been able to move (run and jump) as freely and actively as a healthy child does.

In some cases in which the motor defect is more marked, although the foot is raised from the ground the patient is unable to advance it; and instead of progressing forwards he may go slightly backwards at each step, owing to the foot, after being raised from the ground, falling back further than it was before. Patients affected with pseudo-hypertrophic paralysis are usually unable to seat themselves slowly and steadily down on a chair; they go down suddenly, all of a heap.

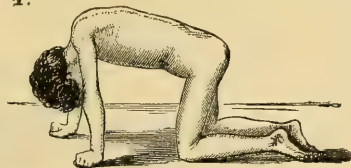
The attitude and gait which I have just described may be absent or very slightly marked in those cases in which the hypertrophy of the calf muscles is absent or slight. I have at present under my care in hospital a very interesting and important case of this kind, the details of which are mentioned below (see page 262).

Peculiar method of rising from the recumbent position; climbing up the thighs.—Another remarkable peculiarity which results from the muscular weakness is the great difficulty which the patient experiences in rising from the recumbent and sitting postures. Indeed, in advanced cases he may not be able to raise himself at all. Even in the earlier stages of the disease, he makes use of his arms, catching hold of chairs or other pieces of furniture and drawing up his body by the aid of the upper extremities.

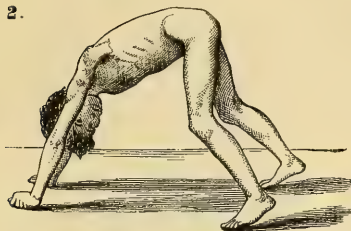
When he is laid flat on his back on the ground and made to rise, without having anything to take hold of, he usually goes

through a series of movements which are represented in figs. 70, 71, and 72. In many cases, he first turns on his face; he then gets

1.



2.



3.

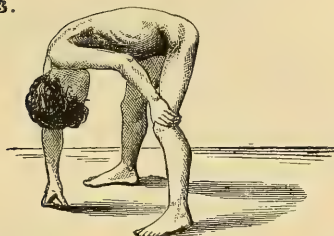


FIG. 70.—*The attitudes which patients with pseudo-hypertrophic paralysis assume in rising from the recumbent to the erect position. —(After Gowers.)*



FIG. 71.—*Climbing up the thighs in pseudo-hypertrophic paralysis. —(After Gowers.)*

with great difficulty on to his hands, the head hanging down between the arms. From this position, he gradually extends

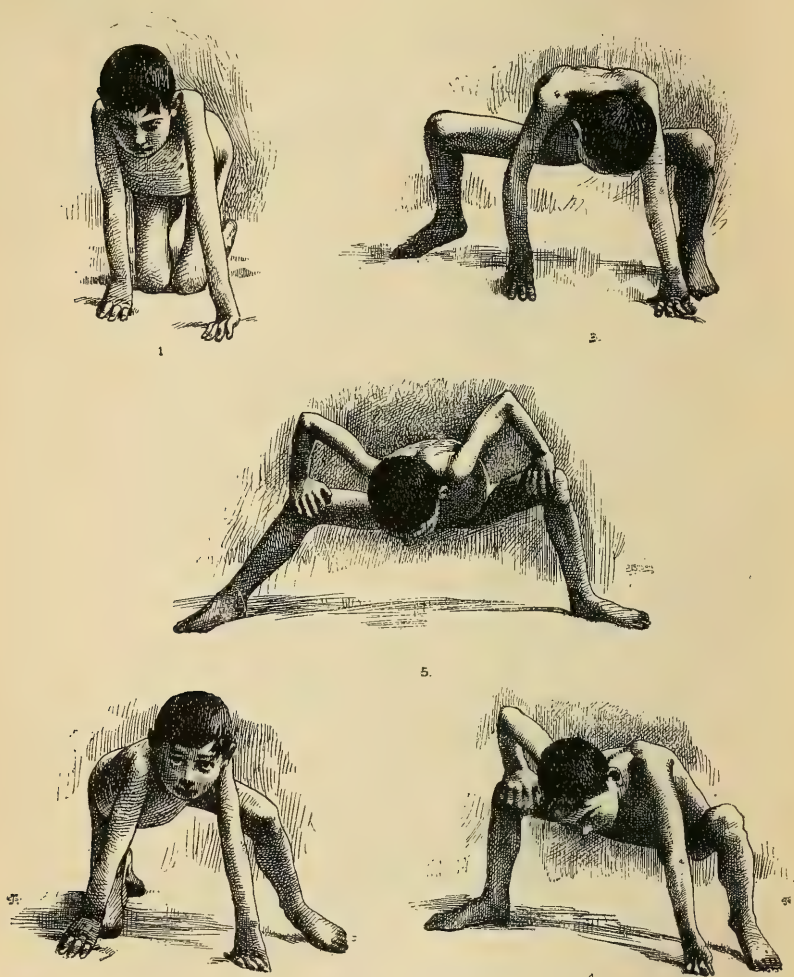


FIG. 72.—Method of rising from the recumbent to the erect position in the case of pseudo-hypertrophic paralysis represented in figs. 73 and 74.

himself, first raising one leg and then the other, and finally 'climbs up his thighs,' as it has been termed. The hip joint is extended by grasping the thigh with the hand and the body is pushed up, as it were, by the arm.

In some cases, the feet are placed close together before the patient raises himself to the erect position. In one of my cases, after the patient had raised himself on to his toes and fingers, the foot and leg were drawn forward by means of the hand (see figs. 84 and 85, page 253). In another, the legs were always placed extra-

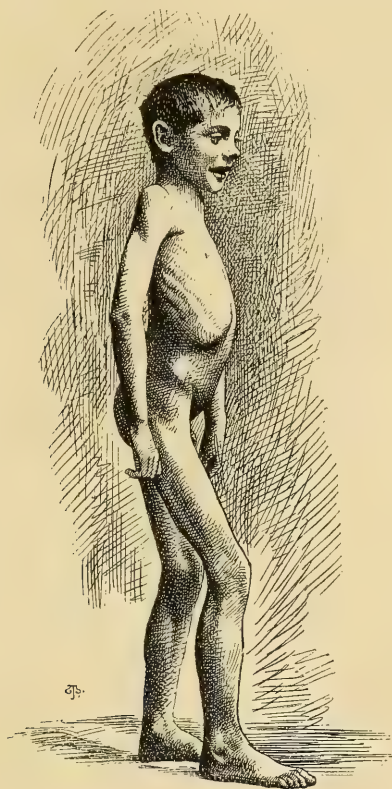


FIG. 73.—*Pseudo-hypertrophic Paralysis.*

The calf muscles are relatively enlarged; at a former stage of the case they were greatly enlarged. The muscles of the thighs and upper extremities are extremely atrophied. The patient walked with a 'spider-crab gait.'

ordinarily wide apart (see fig. 72). It was a remarkable proof of the patient's co-ordinating power that he was able to raise himself with a jerk from this into the erect position. When this patient came under my notice there was little or no muscular enlargement (see figs. 73 and 74) and the case might easily have been placed under the atrophic form (see page 262); but the calves,

which were still relatively large, had at a previous stage of the case been very much enlarged.

The peculiar manner in which the patient rises from the ground and climbs up his thighs is very characteristic, though not pathognomonic. It may occur in any condition in which

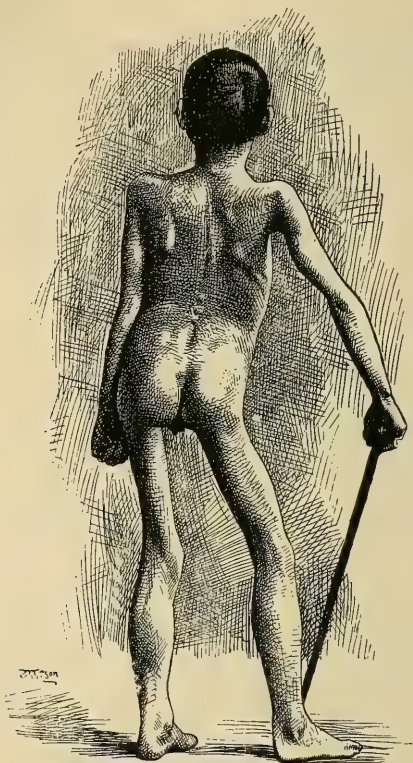


FIG. 74.—*The patient represented in fig. 73.*

there is great weakness of the extensor muscles of the hip. I have seen it in children affected with rickets and in an adult woman who suffered from a diseased (malacostial) pelvis.

Even at an early stage of the disease, the patient experiences difficulty in going upstairs. This and the high action gait are due to the fact that the muscles which flex the thigh on the abdomen are generally affected in an early stage of the case.

Should the patient come under observation at an early period

of the disease, the calf muscles will probably be found to be large, firm and elastic; in many cases they feel like hard masses of india-rubber. The enlarged muscles stand out prominently, as if they were in a permanent condition of contraction; but notwithstanding this apparent hypertrophy their motor power is (usually) markedly impaired. The amount of contraction, too,

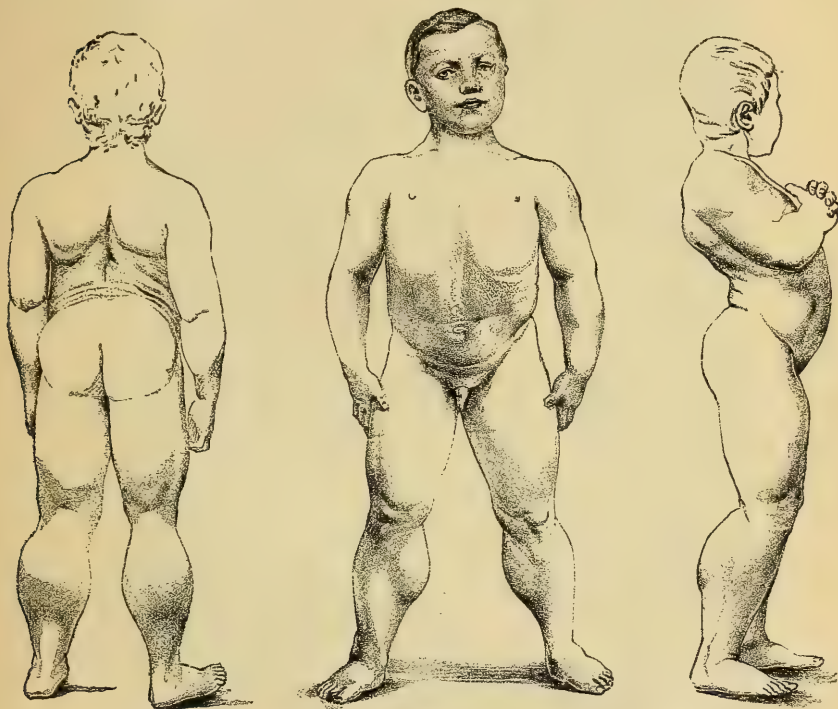


FIG. 76.

FIG. 75.

FIG. 77.

Fig. 75, front view; Fig. 76, back view; and Fig. 77, side view of a case of pseudo-hypertrophic paralysis, in which almost all the muscles of the body were hypertrophied.—(After Duchenne.)

which can be produced in them by a powerful faradic current is below the normal.

In some cases, the gluteal muscles or the muscles on the front of the thigh are also enlarged. The deltoids and infraspinati are in many cases markedly enlarged, apparently hypertrophied. The enlargement of the infraspinati was very remarkable in the case which is represented in figs. 64 and 65.

In rare cases (see figs. 75, 76, 77), almost all the voluntary

muscles of the body may become enlarged. This, however, is quite exceptional.

In the great majority of cases, muscular atrophy is a prominent feature even in the earlier stages of the disease. The

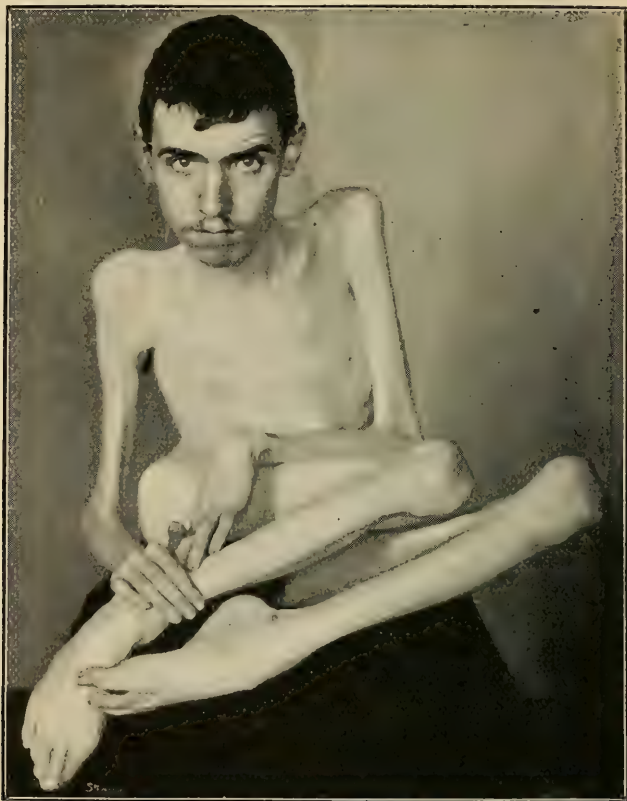


FIG. 78.—*The last stage of pseudo-hypertrophic paralysis, showing extreme atrophy of the muscles and contractures.*

latissimus dorsi and the lower portion of the pectoralis major are almost invariably wasted. As Gowers has pointed out, this is a point of great diagnostic importance.

The small muscles of the hand (fingers and thumb) are very rarely affected in the early stages of the disease; but they may be atrophied in the terminal stages. The muscles of the face

and neck are very rarely indeed affected in a marked degree, and it is only in exceptional cases that the tongue is involved.

In the later stages, the hypertrophied muscles become atrophied; and in the course of years, provided the patient lives long enough, almost all the voluntary muscles of the body



FIG. 79.—*The same patient seen from behind, showing a marked spinal curvature.*

(the intrinsic muscles of the hand and the muscles of the face and neck usually excepted) may appear to have completely disappeared.

The case which is represented in figs. 78 and 79 shows the last stage of the disease. The history of the case is briefly as follows:—

The patient was one of a family of seven, three boys and four girls. All the boys have been affected, all the girls have escaped. The eldest boy died at the age of 18 $\frac{3}{4}$, shortly before I saw his two younger brothers. For some years previous to his death, his condition was exactly similar to that of the patient.

The second brother (see figs. 78 and 79) began to suffer from the disease about the age of six. When he came under my notice (he is since dead, and in his case the spinal cord was malformed as shown in fig. 60) he was reduced to the condition of a helpless cripple; the muscles of the limbs and trunk had almost entirely disappeared; the patient looked like a skeleton; the muscles of the face did not appear to be affected. The tongue was rather large. He died at the age of 18 $\frac{1}{2}$.

The third brother, a boy aged 8 $\frac{1}{2}$ years (see fig. 80) presented all the characteristic features of the disease in its early stage when he first came under my notice; the characteristic attitude, gait and peculiar method of rising from the recumbent position were quite typical, and the calf muscles were notably enlarged. In his case, as in the case of his brothers, the disease has advanced with great rapidity, and now at the age of ten he (like his brothers at the same age) has, in the course of a few months, lost the power of standing and walking.

The four girls, aged respectively when I first saw them, 16, 14, 12 and 10 years, have escaped. The eldest is tall and rather delicate; the three younger girls are well-grown, stout, active and strong.

The father and mother are healthy people and, so far as is known, no cases of the disease have occurred in any other of the near relations.

In the advanced stages of the disease, the spinal column is often markedly curved (see fig. 79).

When the latissimus dorsi and pectoralis muscles are weak and atrophied, the shoulders offer no resistance but are pushed up towards the ears when an attempt is made to raise the patient into the air, the hands being placed under the arm-pits (see fig. 81). Erb lays stress upon this and also upon the fact that if the arm, raised to the horizontal level, is brought forcibly down against powerful opposition, the scapula is drawn strongly towards the upper arm and its angle is moved outwards, in consequence of defective fixation by the trapezius and rhomboids.



FIG. 80.—*Early stage of pseudo-hypertrophic paralysis. The patient is a younger brother of the patient represented in figs. 78 and 79.*

The calf muscles are enlarged. The gait and method of rising from the recumbent to the erect position were quite characteristic.

The photograph was taken when the patient was $8\frac{1}{2}$ years old. At the age of 10 the disease had made such rapid progress that the patient was unable to stand or walk.



FIG. 81.—Case of progressive muscular dystrophy (the case represented in fig. 87 and described on page 262), showing the inability to fix the scapulae and shoulders to the trunk.

When the photograph was taken the patient was trying to hold the shoulders down.

The inability to fix the scapula to the thorax is well seen in fig. 82.

The condition of the muscles.—As I have already told you, some of the muscles appear to be hypertrophied, while others are atrophied. As the disease advances, the enlarged muscles waste. Fibrillary twitchings are very rarely indeed present. Erb says very emphatically that fibrillary tremors or twitchings are never present. With this opinion I am unable to agree. In

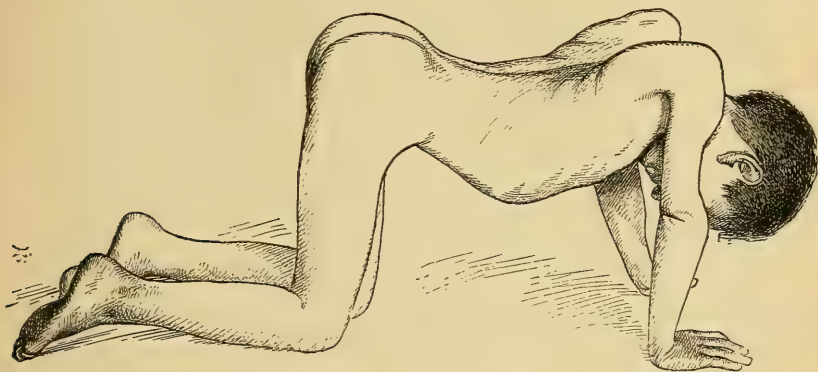


FIG. 82.—Case of *pseudo-hypertrophic paralysis* (the case represented in figs. 63 and 64) showing the inability to fix the shoulder blades to the trunk.

The photograph was taken when the patient was attempting to rise from the recumbent to the erect position.

the case which is represented in figs. 81 and 87, fibrillary twitchings were very conspicuous in the muscles of the back, which were very markedly atrophied; they were also occasionally noticed in some of the muscles of the limbs. The case is of great interest not only because of the presence of fibrillary twitchings but because it conclusively shows the identity of pseudo-hypertrophic paralysis with the atrophic form of progressive muscular atrophy.

The more important particulars of the disease in this family are as follows:—

The patient, a girl aged 11, is one of six (surviving) children—two boys and four girls. Two of the boys and two of the girls are affected. The ages are as follows:

1. Male, aged $13\frac{2}{12}$, the pseudo-hypertrophic form. In his case the disease commenced at the age of 14 months; it has progressed slowly; he is still able to walk and go to school.

2. Female, aged 11, the patient represented in figs. 81 and 87. In her case the disease commenced at the age of 3 years; it has advanced rapidly. In this case there is no, and there has never been any, muscular enlargement, unless the gluteal muscles are hypertrophied (the buttocks look relatively, to the other parts of the body, large, but not larger than they might quite well be in a healthy girl of the patient's age and size). The muscles of the limbs, including the small muscles of the hands and back are markedly atrophied.
3. Female, aged 10, unaffected.
4. Male, aged 8; the pseudo-hypertrophic form. In this case the disease commenced at the age of 6 years, and is still in an early stage.
5. Female, aged 6; affected. In this case the disease is in a very early stage; it commenced at the age of 5 years; the calves are enlarged, and the disease promises to assume the ordinary pseudo-hypertrophic form.
6. Female, died at the age of 5 months.
7. Female, aged $1\frac{1}{2}$ years; as yet unaffected.

In cases of pseudo-hypertrophic paralysis, the wasted muscles respond feebly to electric currents (simple diminution), but the reaction of degeneration is rarely, if ever, developed. The faradic irritability diminishes as the muscular atrophy increases; in the later stages the atrophied muscles may fail to respond to either form of current.

The mechanical irritability is also diminished.

In the advanced stages of the disease, contractures at the ankle, knee and elbow, less frequently at the hip, are often produced (see fig. 78).

The condition of the reflexes.—In the earlier stages of the disease, the reflexes may be unaltered; but in those cases in which the muscles on the front of the thigh are markedly affected, the knee-jerks may be absent at a comparatively early period of the case. The skin reflexes are usually preserved for a long time.

The functions of the bladder and rectum are seldom interfered with, but occasionally a temporary condition of incontinence occurs in the later stages of the disease.

Provided that the patient attains the age of puberty, the sexual development takes place in the normal manner. In some cases it is precocious.

Vasomotor and trophic alterations.—The nutrition of the bones does not seem to be interfered with. The osseous development goes on. In the case which is represented in figs. 78 and 79, the bones seemed to have developed, as regards their length at least, quite normally.

The skin of the legs and thighs often has a mottled appearance; this is not constant and is by no means pathognomonic. I have noticed exactly the same appearance in several other conditions, notably in the spastic paraplegia of early life. In some cases, the feet and hands are blue and cold. The surface temperature is in some cases subnormal.

Sensory functions.—The sensory functions of the skin and the special senses are always quite normal.

Mental condition.—The intellectual faculties and cerebral functions are, in many cases, absolutely normal. Some of the patients affected with pseudo-hypertrophic paralysis are intellectually above the average. The patient represented in fig. 78 was mentally very acute; he employed his time in reading all sorts of literature. In other cases, the mental faculties are imperfectly developed; occasionally the patient is quite idiotic. The patient who is represented in fig. 86 is intellectually very much below the average; his brother, who is also affected, is mentally very deficient; and his sister, who presents none of the features of the disease, is deranged and in an asylum.

Course and Duration.—The duration of pseudo-hypertrophic paralysis varies very considerably in different cases. The disease generally runs a very chronic course. Cases which develop soon after birth seem, as a rule, to progress more rapidly than those which are developed about the age of puberty. This fact is perhaps due to the circumstance that the tendency to the disease is born with the individual, and that the stronger the congenital tendency, the earlier is the disease likely to manifest itself and the more severe is it likely to be. Girls, as I have previously remarked, are usually attacked later than boys; and in girls the disease is usually less severe than in boys.

When the disease does not develop until the period of puberty, the prognosis, as regards duration, is much more favourable.

Death is usually due to the occurrence of some pulmonary

complication (such as bronchitis or pneumonia) or other intercurrent disease. A comparatively trivial illness may easily kill when the patient is reduced to the condition of the boy represented in fig. 79; he died from influenza and bronchitis.

Diagnosis.—In typical cases the clinical picture is so striking that the diagnosis is self-evident, provided of course that the observer has seen a case of pseudo-hypertrophic paralysis before. The attitude and gait, and the peculiar way in which the patient raises himself from the ground by climbing up his thighs (though this is not absolutely pathognomonic) are highly characteristic. The enlargement of the calf muscles is, however, the most striking and important feature of this variety of myopathic muscular atrophy from a diagnostic point of view, for in some of the other forms the same attitude, gait, and mode of rising from the recumbent position may be present. The enlargement of the infraspinati muscles is also of great diagnostic significance. The absence of fibrillary twitchings and the reaction of degeneration are, as I have more than once stated, characteristic features of this and the other forms of myopathic muscular atrophy. The age and sex of the patient are of diagnostic value; and the fact that several members, usually males, in the same family are often affected is very significant. Gowers lays much stress upon the atrophy of the lower part of the latissimus dorsi and pectoralis major muscles.

It is important to remember that the muscular atrophy is in some cases associated with very little or no pseudo-hypertrophy (see figs. 73 and 81). Unless the observer is well acquainted with the various forms of myopathic muscular atrophy, the true nature of cases of this kind may be easily overlooked, especially if the case is isolated, i.e. when no other members of the family are affected.

To sum up, the most important clinical features of pseudo-hypertrophic paralysis from a diagnostic point of view are:—The slow and gradual onset; the affection of several members, usually boys, in the same family; the gradual and progressive muscular weakness; the enlargement of certain muscles, especially the calf muscles, glutei, infraspinati and deltoids; the atrophy of other muscles, especially the lower part of the latissimus dorsi and pectoralis major; the absence of fibrillary

twitchings and of the reaction of degeneration; the peculiar attitude and gait, and the characteristic mode of rising from the recumbent to the erect position; the inability to fix the scapulæ to the thorax; the normal, diminished, or abolished (never exaggerated) condition of the knee-jerks; the normal condition of the bladder and rectal reflexes; and the entire absence of sensory symptoms and of visceral lesions.

The differential diagnosis of pseudo-hypertrophic paralysis and of Friedreich's ataxia.—These diseases have certain features in common. Both are diseases of development (i.e. they very rarely commence after the development of the body is completed); both are 'family diseases' (i.e. they are apt to affect several members of the same family); both produce difficulty in walking; in both, there is muscular weakness; in both, the knee-jerks may be abolished (almost always in Friedreich's ataxia, often in pseudo-hypertrophic paralysis); in both, the spine becomes curved in the later stages; in both, the feet may become distorted (clubbed); and in both, the patient is finally reduced to the condition of a helpless cripple.

But, notwithstanding these points of resemblance, there is no difficulty in distinguishing the two conditions in the earlier stages of the case. In Friedreich's ataxia, the difficulty in walking is due to inco-ordination rather than to pure muscular debility. In pseudo-hypertrophic paralysis, on the other hand, the difficulty in walking is entirely the result of muscular weakness, the co-ordinating power being retained in a very remarkable way. The attitude and gait are quite different in the two diseases. In Friedreich's ataxia, the hands are affected as well as the lower extremities; speech, too, is often involved; whereas, in pseudo-hypertrophic paralysis, the movements of the hands are rarely interfered with even in the later stages, and speech is never affected. The enlargement (apparent hypertrophy) of the muscles which is so characteristic of pseudo-hypertrophic paralysis is never seen in Friedreich's ataxia. The atrophy of the lower part of the latissimus dorsi and pectoralis major, and it may be of other muscles, is also an important point of difference.

The differential diagnosis of pseudo-hypertrophic paralysis and the spastic paraplegia of early life presents no difficulty. The chief points of distinction are:—

The mode of onset. Slow and gradual in pseudo-hypertrophic

paralysis; often rapid as the result of a distinct cerebral lesion, associated with convulsions, or due to an injury received at the time of birth (instrumental delivery) in the spastic paraplegia of infancy.

The condition of the deep reflexes. Normal, diminished or abolished in pseudo-hypertrophic paralysis, exaggerated in the spastic paraplegia of early life.

The condition of the upper extremities. In the former, some of the muscles of the shoulder girdle (especially the lower part of the pectoralis major and the latissimus dorsi) are usually atrophied and others (especially the deltoid and infraspinati) are hypertrophied; while, in the latter, all of the muscles of the upper extremity may be more or less atrophied and undeveloped, or rigid with exaggeration of the deep reflexes.

The attitude and gait. The lordosis, waddling, high-action gait, and peculiar mode of rising from the recumbent to the erect position which are such characteristic features of pseudo-hypertrophic paralysis, are all absent in the spastic paraplegia of early life. In the latter condition, the legs are more or less rigid; when the patient stands or is held in the erect position, the feet tend to cross and to assume the position shown in fig. 83.

The presence of cerebral symptoms. Although in some cases of pseudo-hypertrophic paralysis the intellectual development is interfered with, this is exceptional; many of the patients are mentally very acute. But in patients affected with spastic paraplegia dating from early life, there is usually some mental impairment, and recurring epileptiform attacks are often present.

The hereditary and family tendency. In cases of spastic paraplegia of early life, the hereditary and family tendency, which is such a conspicuous feature of most cases of pseudo-hypertrophic paralysis, is entirely absent.

Prognosis.—The prognosis of pseudo-hypertrophic paralysis is very unfavourable. The tendency of the disease is to progress steadily from bad to worse; almost all cases terminate, sooner or later, in death; but in many cases the fatal issue is not reached for several years. When the extreme condition of emaciation which is represented in figs. 78 and 79 is reached, the patient may still live for several years, but his tenure of life is very insecure. A slight bronchial attack, or any intercurrent

respiratory or other disease may in such circumstances speedily prove fatal. Speaking generally, the earlier the disease is developed, the more rapid its course; and since the disease usually develops earlier in males than in females, the prognosis as regards the rapidity is worse in boys than in girls.



FIG. 83.—Case of infantile spastic paraplegia (bilateral hemiplegia) dating from the time of birth; showing the attitude and position of the limbs when the patient was supported in the erect position.

Treatment.—The treatment which has hitherto been employed has been of little avail. We know of no remedy which exerts any distinctly beneficial influence upon the course of the disease. The general health must be kept in the best possible state of efficiency. The patient should be well fed, well clothed and have plenty of fresh air. General tonics, more especially cod-liver oil, are advisable. The nutritive condition of the

affected muscles should be, so far as possible, maintained by judicious massage, suitable gymnastics, systematic muscular exercises and the faradic current. Duchenne states that he cured two cases of the disease by the interrupted current; and Erb thinks that electrical treatment is the most important means of influencing the disease which we possess. He advises that the galvanic current should be applied to the spine over the lumbar and cervical enlargements of the cord, with the object of acting upon the trophic centres situated in these regions; and that the faradic, galvanic, or farado-galvanic currents should be applied directly to the muscles and nerves. Strong currents and long applications must not, however, be employed.

Gowers has pointed out that when, from any cause, the patient ceases to walk, the weakness in the muscles is apt to increase rapidly. Contractures and deformities which interfere with walking should therefore, if possible, be prevented by passive exercise and section of the tendons. The application of mechanical appliances and supports has also been recommended. A tight-fitting belt or jacket in some cases seems to be helpful; but mechanical supports (irons, etc.) for the limbs are, in my opinion, useless or injurious.

The patient should be carefully guarded against cold and exposure; for in this, as in all forms of chronic spinal disease, cold seems to exert a depressing influence upon the condition.

Amongst drug remedies, arsenic and strychnine are probably the most useful. The systematic use of hypodermic injections of strychnine deserves, I think, a much more thorough and extended trial in this disease than it has hitherto received.

It is important to note that in some cases of pseudo-hypertrophic paralysis some temporary improvement may undoubtedly be effected by judicious treatment. Under arsenic, strychnine and the faradic current, the patient represented in fig. 87 undoubtedly improved. At the time of her admission to the hospital she was quite unable to raise herself from the recumbent position; she could get into a sitting position and that was all. After two months' treatment, she could raise herself to the erect position with comparatively little effort or difficulty.

The internal administration of extract of thymus gland has been recommended in pseudo-hypertrophic paralysis. In the few cases in which I have prescribed it, I cannot say that it has

produced any beneficial effect. Two of the patients have undoubtedly improved, probably I think in consequence of the arsenic, strychnine, massage and electricity which have also been employed.

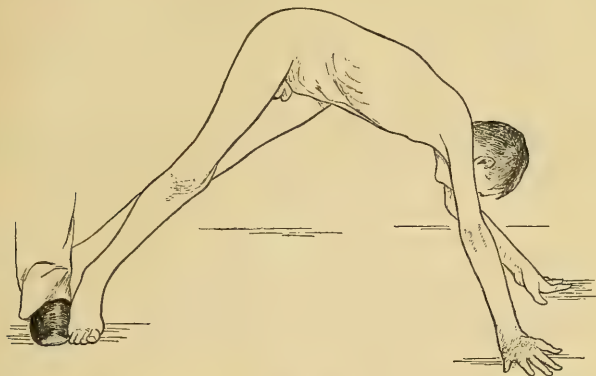


FIG. 84.

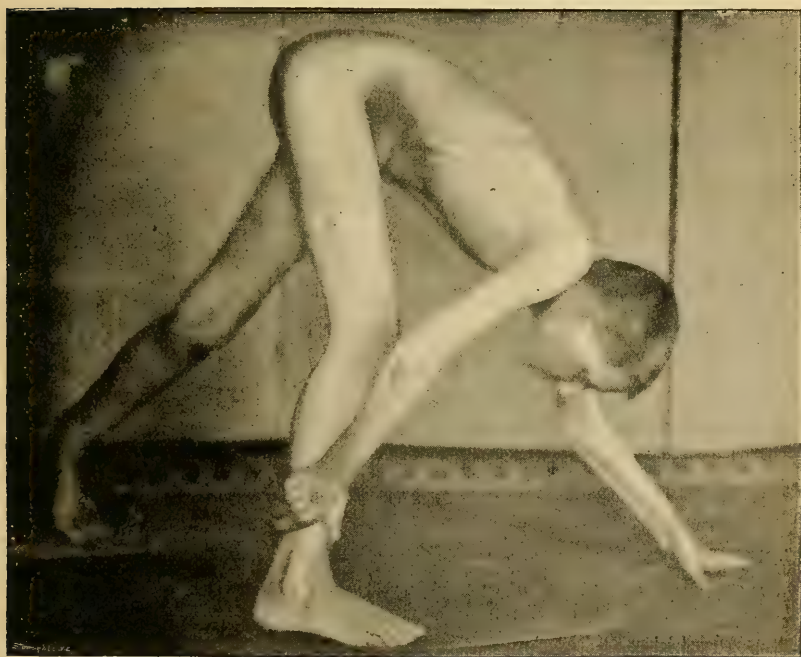


FIG. 85.

FIGS. 84 and 85.—*Peculiar method of rising in a case of pseudo-hypertrophic paralysis.*

LECTURE XV

THE JUVENILE FORM OF MYOPATHIC MUSCULAR ATROPHY OR PROGRESSIVE MUSCULAR DYSTROPHY OF ERB

ALTHOUGH the juvenile form of progressive muscular atrophy usually commences in childhood or early youth, the average age at which it is developed appears to be somewhat later than that at which pseudo-hypertrophic paralysis commences. This form seems to begin much more frequently in later youth and early adult life than the pseudo-hypertrophic form. Females, too, are more frequently affected than in the pseudo-hypertrophic form.

The disease seems to run a slower and less active course than most cases of typical pseudo-hypertrophic paralysis. Many cases live for a long time, and some have been reported in which the patients have attained old age. In short, the tendency to kill is not so strong as in the pseudo-hypertrophic form.

In this form, as in pseudo-hypertrophic paralysis, the hereditary and family tendency is very strong.

The disease is characterised by a slow and gradually developed muscular weakness and atrophy, which in the great majority of cases tends to be progressive; and which is associated with enlargement, due either to a true or false hypertrophy, of certain muscles. But in the order of the development and the distribution both of the atrophy and the hypertrophy it differs from the more common pseudo-hypertrophic type. In typical cases of pseudo-hypertrophic paralysis, the weakness and atrophy commence in the legs and back, and the calf muscles are enlarged from pseudo-hypertrophy (lipomatosis). In Erb's juvenile form, the atrophy and muscular enlargement, in the earlier stages of the case at all events, chiefly involve the muscles of the shoulder and pelvic girdles.

In the upper extremity, the pectoralis major and minor

(the clavicular portion usually excepted), the latissimus dorsi, trapezius, serratus magnus, rhomboids, biceps, brachialis anticus and supinator longus are usually atrophied; while the deltoids, infraspinati and triceps are usually enlarged, often apparently as the result of an actual hypertrophy. The muscles of the forearm (with the exception of the supinator longus) and hand usually escape or remain unaffected until the later stages of the disease.

According to Erb, well-preserved forearms, atrophied upper arms, hypertrophied deltoids and atrophied scapular muscles (the infraspinati excepted) are characteristic of this type.

In the lower extremity, the muscles of the hip and thigh, especially the glutei, the flexors of the hip and the extensors of the knee are usually atrophied. In the later stages, the muscles both on the front and back of the leg are often involved; but the calf muscles seem to be much less frequently affected during the earlier periods of the disease than in typical pseudo-hypertrophic paralysis.

In many cases, the muscles of the back (sacro-lumbalis and longissimus dorsi) are involved even in the early stage.

In rare cases, the facial muscles (chin and cheek muscles especially) are involved. The affection of the muscles of the face brings this form into relationship with the next type—the facio-scapulo-humeral type of Landouzy and Déjerine.

As a result of the muscular atrophy and wasting, deformities and lordosis are developed.

As in pseudo-hypertrophic paralysis, fibrillary twitchings are absent¹ and the reaction of degeneration is not developed.

The knee-jerk may be normal, diminished or lost, but is never exaggerated. There are no sensory symptoms; no affection of the bladder and rectum; no visceral lesions. Further, as I have already so often remarked, our present methods of investigation fail to show any lesion either in the cord or in the nerves.

From this description, you will see that the disease is closely allied to pseudo-hypertrophic paralysis. There can, I think, be little doubt that the two diseases are mere variations of one primary form. I agree with Erb in thinking that the differences which the two types present, as regards the distribution of the atrophy and hypertrophy, the order of development and the

¹ On this point, see page 245.

varying degrees of intensity of the muscular lesions, are not of themselves sufficient to prove that the two conditions are fundamentally distinct. (See page 211.) The case represented in fig. 86 may perhaps be placed under this type. The clinical features were briefly as follows:—

The patient (see fig. 86), a boy aged 16, is one of a family of 9 children. One died at the age of three months from (the mother says) teething. Of the 8 who are alive, 4 are boys and 4 girls. Of the 4 boys, two are affected. One girl is insane, and both the boys affected with pseudo-hypertrophic paralysis are weak-minded. The other members of the family are all very bright and sharp.

In the patient's case (fig. 86), the back is markedly arched, the calves are relatively enlarged, the thighs and buttocks are normal, the deltoids are distinctly enlarged, the infraspinati and scapular muscles are atrophied, the pectoralis major is markedly atrophied, the muscles of the upper arms are moderately atrophied, the muscles of the forearm and hands are normal.

The knee-jerks are present but greatly diminished in degree.

The arching of the back (dorso-lumbar curve and throwing back of the shoulders) are the only characteristic alterations in the gait.

The patient's brother who is also affected has suffered all his life from nocturnal incontinence of urine.

The shoulders go up to the ears (as in the case represented in fig. 81). The patient gets up from the ground in the characteristic pseudo-hypertrophic fashion. The hands are blue and cold (vasomotor derangement). The mental condition is much below the average.

As Sachs has pointed out, the juvenile form of Erb is much less frequently met with than the typical pseudo-hypertrophic form.

The later age at which the disease usually commences, its longer duration, its lesser tendency to kill and the greater frequency with which females are affected may all probably be explained by the lessened intensity which the disease presents. I have already pointed out that when pseudo-hypertrophic paralysis develops late, it is apt to be less severe; and that, in the female, the disease usually develops later and is less severe than in the male.

Diagnosis.—The diagnosis of the juvenile form of progressive muscular atrophy is usually easy, provided that the observer is

acquainted with the characters which this and the other forms of myopathic atrophy present. The fact that several members



FIG. 86.



FIG. 87.

FIG. 86.—Case of *progressive muscular dystrophy* (? Leyden's form).

FIG. 87.—Case of *idiopathic muscular atrophy* (*progressive muscular dystrophy of Erb*).
The chief clinical features of the case are described on page 262.

of the same family (brothers and sisters of the patient) are affected, or that other cases of a similar or allied nature have occurred amongst the patient's relatives (in his cousins, uncles, aunts, grandparents, etc.) is of great diagnostic importance.

The combination of atrophy and hypertrophy, and especially the peculiar distribution which the atrophy and hypertrophy present—well-preserved hand and forearm muscles, atrophied upper arms, enlarged deltoids and infraspinati and atrophied scapular muscles—are highly characteristic. The absence of fibrillary tremors and of the reaction of degeneration, diminished mechanical irritability and diminution or abolition of the deep reflexes are also of great importance.

From the pseudo-hypertrophic form, Erb's juvenile type is distinguished (though it must be remembered that this is not constant for in the juvenile form the calves may also be enlarged) by the absence of enlargement of the calf muscles, by the fact that the upper extremity is first affected, and by the peculiar way in which the atrophy and hypertrophy are distributed in the muscles of the shoulder girdle and upper limb. But I repeat that the two diseases seem to run insensibly one into the other; and that connecting types and irregular forms occasionally occur.

Prognosis.—Speaking generally, this seems to be more favourable than in the pseudo-hypertrophic form. Different cases differ, however, very markedly as regards their severity. In some, the course is rapid; in others, very chronic. As I have already said, some patients live on and reach old age. In others, the atrophic process seems to be arrested after a certain stage; but it must be remembered that in such cases redevelopment may occur after an interval of quiescence and apparent arrest.

In trying to form an opinion as to the severity in any particular case, the age at which the disease develops, the rapidity with which the atrophic process is progressing, the number of muscles which are affected, and especially the condition of the respiratory muscles and of the diaphragm are the most important points.

In those cases in which the disease is developed early the course is usually more rapid and severe than in those in which the disease commences later.

Treatment.—The same methods of local and general treatment which have been recommended for the treatment of pseudo-hypertrophic paralysis are advisable.

THE FACIO-SCAPULO-HUMERAL TYPE OF LANDOUZY AND DÉJERINE.

This is identical with the infantile form of progressive muscular atrophy of Duchenne. As I have already stated, it seems to be merely a modification of Erb's juvenile form and of pseudo-hypertrophic paralysis. Its peculiarity consists in the fact that the facial muscles are affected, and that they are (usually) the first muscles which are involved.

In this country, this type of the disease seems to be extremely rare. One case only has come under my own observation. This form also seems to be very rare in Germany, for Erb with his immense experience does not seem to have met with a single typical example of it. It would appear to be much more common in France.

In this form, the disease usually develops in childhood or early youth. Like the other forms of myopathic atrophy, it presents a strong hereditary and family tendency.

The affection of the facial muscles, when advanced, gives a peculiar appearance to the patient which has been termed the *facies myopathique*. The face is flattened, dull and expressionless; the lips, more particularly the lower lip, appear to be enlarged; the lower lip is everted (pouting); the mouth has been compared to that of a tapir (*bouche de tapir*). When the patient smiles, the angles of the mouth are not raised, but the mouth is drawn out transversely (*rire en travers*). The pronunciation of labials is more or less interfered with. The patient is unable to whistle (see fig. 88). When the disease is advanced, the wrinkles on the forehead may be completely effaced; in some cases, the orbicularis palpebrarum is so markedly affected that the eyes cannot be closed even during sleep.

In addition to the affection of the facial muscles, some of the muscles of the shoulder girdle and of the pelvic girdle are usually affected, as in Erb's juvenile form. Ultimately, almost all the muscles of the body may become involved.

Diagnosis.—When the affection of the face is advanced, this type is easily recognised by the peculiar expression, the character of the smile and the tapir-like form of the mouth. The early age at which the disease is developed is also in most cases an important aid to diagnosis.

When the facial muscles are only affected in a slight degree, the atrophy may easily escape observation. And here I may say that it is of great importance to examine the condition of the facial muscles in all cases of suspected myopathic atrophy (pseudo-hypertrophic paralysis, Erb's juvenile form) with the



FIG. 88.—*Case of progressive muscular dystrophy. The facio-scapulo-humeral.*

Type of Landouzy and Déjerine.

The photograph was taken while the patient was attempting to whistle. The lips cannot be closed.

greatest care, not only during life but also after death. Post-mortem observation has shown that, in some cases in which the face appears to be perfectly normal during life, the facial muscles are actually involved. This (unobtrusive) implication of the

facial muscles in the pseudo-hypertrophic and juvenile forms, speaks strongly in favour of the view that the three types are merely modifications of one and the same disease.

Prognosis.—This seems to be more unfavourable as regards rapidity of progress and duration than in the juvenile form of Erb, but perhaps more favourable than in typical cases of pseudo-hypertrophic paralysis.

Treatment.—The same as for pseudo-hypertrophic paralysis.

THE HEREDITARY FORM OF PROGRESSIVE MUSCULAR ATROPHY OF LEYDEN.

In this form, which is merely a modification of pseudo-hypertrophic paralysis, the weakness and atrophy usually commence in the muscles of the legs and back. In some cases, the calf muscles are enlarged or relatively enlarged in proportion to the atrophied condition of the thighs. Lordosis similar to that which characterises pseudo-hypertrophic paralysis is a striking feature. In some cases, the gait is very similar to that of pseudo-hypertrophic paralysis, but this is by no means always the case; and the same peculiar method of rising from the recumbent to the erect position (climbing up the thighs) is observed.

The family tendency is strongly marked; hence the term, the *hereditary* form of progressive muscular atrophy, which Leyden applied to the disease. Several members of the same family are often affected. Fibrillary tremors and the reaction of degeneration are almost invariably absent, and the small muscles of the hand are usually spared until, at all events, the later stages of the case.

The disease usually develops in childhood or early youth, though generally at a later age than pseudo-hypertrophic paralysis. The course seems, as a rule, slower than in typical cases of pseudo-hypertrophic paralysis, for the intensity of the disease seems less. The case represented in fig. 86 falls under this type.

THE (PURELY) ATROPHIC FORM OF MYOPATHIC MUSCULAR ATROPHY.

Cases are occasionally seen, but they are probably very rare, in which progressive muscular weakness and atrophy are developed, and in which there is no muscular hypertrophy. The muscles of the back and leg are usually first affected, but ultimately the atrophy may involve almost all the muscles of the body. In these cases, the characteristic lordosis and the same method of rising from the recumbent to the erect position is seen as in the pseudo-hypertrophic form. The gait is in some cases peculiar, but not exactly identical with that of typical pseudo-hypertrophic paralysis; it is not so waddling. I am in the habit of terming it the 'spider-crab' gait.

When the patient is stripped, the great tenuity of the limbs in proportion to their length, is a very striking feature. In particular, the thighs appear to be atrophied; the calf muscles though not actually increased in size, usually, so far as my experience enables me to judge, appear to be relatively large. It is not improbable that in many of these cases (as in that represented in figs. 72 and 73) the calf muscles are actually in some degree enlarged, or that they were enlarged prior to the time when the patient comes under observation.

This type seems to be identical with the form previously described (Leyden's form). The case represented in fig. 87 might be placed either under this form or under the type described by Leyden.

As I have already pointed out, the girl represented in fig. 87 is one of a family of six children, two boys and four girls; two of the boys and two of the girls are affected, and in the other three cases in this family the disease presents the usual features of the pseudo-hypertrophic form. The identity of the hereditary form of progressive muscular atrophy of Leyden and the ordinary pseudo-hypertrophic form is conclusively proved by these cases. In the girl's case, there has never been any enlargement of the muscles, except perhaps of the glutei, for the buttocks look relatively large and are elastic and firm to the feel. Another point of

great interest is that in this girl's case fibrillary tremors in the muscles of the back were very noticeable. In another case, which was sent to me in the year 1892 by the late Dr. Milne Chapman of Inverness and which is now under the care of Dr. Forsyth, definite and distinct fibrillary twitchings were also seen. The patient was a boy aged 10. When he was first seen there was no distinct pseudo-hypertrophy. The deltoids and thigh muscles are now markedly enlarged and the facial muscles notably affected (see fig. 88).

Diagnosis.—When cases of this kind occur in an isolated form, the difficulty of diagnosis may be very great, unless the observer is well acquainted with the modifications which myopathic atrophy may present. These cases seem to be a mere modification of the pseudo-hypertrophic form. They are pseudo-hypertrophic paralysis without pseudo-hypertrophy.

The differential diagnosis of the purely atrophic form of myopathic atrophy and of the myelopathic (Aran-Duchenne) type of progressive muscular atrophy.—From the Aran-Duchenne type of progressive muscular atrophy; these cases are distinguished by:—The early age at which the disease develops; the fact that several members of the same family are usually affected, either with this or one of the allied forms of myopathic atrophy; the hereditary tendency which the disease usually presents; the absence of fibrillary tremors and of the reaction of degeneration; the diminution or abolition (never exaggeration) of the knee-jerks; the lordosis; the peculiar method of rising from the recumbent to the erect position; and the distribution of the atrophy and its order of development, for the small muscles of the hand are rarely if ever involved until the later stages of the case.

The differential diagnosis of the purely atrophic form of myopathic atrophy and the peroneal type of the disease.—These two forms can usually be differentiated without difficulty. As I shall presently point out, the peroneal type of the disease first affects the foot and leg muscles, and then, after (usually) a considerable interval of time, extends to the small muscles of the hand, producing the characteristic *clawed* appearance.

The differential diagnosis of the purely atrophic form of myopathic muscular atrophy and the generalised progressive muscular atrophy of early childhood.—These conditions present many points of

similarity, but I doubt whether they are the same disease. So far as my experience enables me to judge, the condition which I term the generalised progressive muscular atrophy of early childhood differs from the purely atrophic form of myopathic atrophy inasmuch as :—(a) it commences at an earlier age ; (b) the atrophy is much more generalised (diffused) ; (c) the course is more rapid ; ?(d) fibrillary twitchings and the reaction of degeneration are usually present ; and (e) the nerve cells in the anterior cornua of the spinal cord and the peripheral nerves (if the case reported by Drs. Thomson and Bruce may be taken as a type) are distinctly affected (atrophied and degenerated).

The Course, Prognosis and Treatment are the same as in pseudo-hypertrophic paralysis.

GENERALISED PROGRESSIVE MUSCULAR ATROPHY OF EARLY CHILDHOOD.

Cases are occasionally met with in which a generalised (diffuse) muscular atrophy, which has not the distribution of that which is characteristic of any of the preceding types and which is unassociated with any muscular hypertrophy or pseudo-hypertrophy, is developed in early childhood. These cases ought, I think, to be placed in a separate group. They would appear to be very rare. They seem to me to be very closely allied to, probably identical with, the subacute inflammation of the anterior cornual region described by Duchenne under the term *paralysie générale spinale antérieure subaiguë*.

Through the kindness of Dr. John Thomson, I had the opportunity of seeing a characteristic example which he has described, with full pathological report by Dr. Bruce.¹ Another case came under my own observation a few months ago.

In Dr. Thomson's case (see figs. 89 and 90) the clinical features were as follows :—

The patient, a young, most intelligent female child, who

¹ *Edinburgh Hospital Reports*, vol. i. page 372.

presented no hereditary tendency to the disease, suffered from profound paralysis and considerable muscular wasting which affected in a varying degree most of the voluntary muscles of the body. The paralysis began insidiously and without known cause, probably when the child was between 12 and 18 months old. The onset of the paralysis was unaccompanied by fits, pain, or febrile disturbance; it seemed to have developed sub-acutely in the lower limbs, which were severely affected for at least a year before the upper ones were affected. After an interval, during which little change was noticed in the lower extremities, the upper limbs and neck became rapidly (within three to four weeks) weakened. The face was never more than very slightly affected.

The wasting developed gradually, but not nearly so rapidly as that of ordinary poliomyelitis anterior acuta. Further, the atrophy was progressive.

In the earlier stages of the case, the muscular weakness seemed out of all proportion to the wasting.

The child lived for five years after the disease developed; during this period, the atrophy continued to advance steadily but very slowly.

The distribution of the wasting and paralysis was quite symmetrical, the lower half of the body being much more severely affected than the upper.

There was never any hypertrophy or pseudo-hypertrophy.

Fibrillary twitchings were seen in some of the muscles, especially in those of the face. Some of the muscles of the lower limbs became slightly shortened (contracted), and the joints distorted (see fig. 90). The muscular co-ordination was unimpaired.

In the earlier stages of the case, the faradic excitability was much less impaired than the galvanic; later, the excitability to both forms of current was more equally affected (diminished); and, finally, the galvanic excitability was more impaired than the faradic. Distinct 'reaction of degeneration' was never found.

After the disease had lasted more than a year and a half, cramp-like pains were complained of. The sensibility to the pain produced by the application of faradic electricity was most curiously deficient. In all other respects the sensibility was normal.

There was never any affection of the bladder or rectum, nor any tendency to the formation of bed sores.

Twenty months before death almost total collapse of the left lung occurred suddenly during an attack of slight bronchitis, evidently favoured by the extreme muscular debility

On post-mortem examination, the muscles presented a variety of appearances which closely corresponded with those described by Erb as characteristic of progressive muscular dystrophy. Some of the muscular fibres were atrophied, others



FIG. 89.—*Dr. John Thomson's case of progressive muscular atrophy in a child.*

The photograph was taken in November 1889. It shows the state of nutrition, the characteristic attitude and the hyperextension of the fingers in the attempt to spread them out.

hypertrophied. In places, there was a great increase of the nuclei of the muscles and of the sarcolemma. Many of the muscular fibres were split in a longitudinal direction; others presented vacuolations. There was some pseudo-lipomatosis.

In the nerves, there were quite a number of round and spindle cells between the fibres. Many of the nerve bundles were compressed and atrophied, and even replaced by fibrous tissue.



FIG. 90.—*Dr. John Thomson's case of progressive muscular atrophy in a child.*

The photograph was taken in May 1892. It shows extreme muscular wasting and distortion of the limbs from contraction of the muscles.

In the spinal cord, a large number of the cells of the anterior horn had entirely disappeared; while the majority of those which remained had undergone atrophy to a greater or less extent.

The case is one of great interest and rarity. It is perhaps difficult to express a decided opinion as to its nature, but I agree with Dr. Bruce in thinking that it was probably myelopathic

rather than myopathic. The following facts seem to support this view:—The onset was more rapid than in most cases of myopathic atrophy; the muscular wasting seemed to be preceded by muscular weakness (paralysis); fibrillary tremors were present; the galvanic excitability was in the later stages much more marked than the faradic; there were definite alterations in the peripheral nerves and in the spinal cord.

It has been suggested that in cases such as this in which the symptoms during life are suggestive or indicative of myopathic atrophy but in which the multipolar nerve cells in the anterior cornua of the spinal cord are atrophied and diminished in number after death, the atrophy and disappearance of the nerve cells is a secondary result of the peripheral muscular wasting. With this opinion I cannot agree. The remarkable case which is represented in figs. 78 and 79 completely disproves it. In that case, the muscles had for years been in an extreme condition of atrophy, but the multipolar nerve cells in the anterior cornua were very numerous and large; their processes were numerous and distinct; the only change which they exhibited was a certain degree of fatty degeneration.

In my own case, the symptoms, mode of onset and course were very similar. The patient was unfortunately only seen once and the case was not exhaustively investigated. Amongst other points, the electrical condition of the muscles was not ascertained. The history of the case is briefly as follows:—

Case of Progressive Muscular Wasting in a Young Child.—J. B., aged $1\frac{1}{2}$, was seen at the Edinburgh Royal Infirmary on March 31st, 1893, suffering from diffuse muscular atrophy.

Previous History.—The patient was a first child; his head was not injured at the time of birth; the labour, though long, was quite natural; instruments were not used.

The child seemed to be perfectly healthy during infancy; he grew and developed well, and nothing was noticed amiss with him until he was nine months old. He then began to lose the power of using his limbs. Up to this date, he seemed to be able to move his limbs as freely as any healthy child. Since the age of nine months, the muscular weakness and inability to move have slowly but gradually increased.

He has never been able to creep, stand or walk. His general health is now, and has always been, quite good. He has never had any illness.

Present condition.—The patient was a well-grown, well-developed child. He had several teeth. The appetite was good. He was a very intelligent child; his mother said that he was mentally more advanced than most children of his age.

The muscles generally were very soft and flabby.

During the whole examination, the patient lay perfectly helpless. He could move his toes and fingers a little, but was unable to draw up his legs or to put his arms to his head. The fingers were pointed, and tended to be hyper-extended. The muscles of the arms and legs were very flabby, soft and markedly wasted. The deltoids in particular were extremely atrophied; and the lower part of the pectoralis major seemed to be wanting. The muscles of the back were also markedly atrophied.

The knee-jerks were absent.

The electrical condition of the muscles was unfortunately not tested. The child was only seen once; he lived at a considerable distance in the country; and although the parents promised to bring him back and have him admitted as an in-patient, they withdrew from this decision and could not be persuaded to bring the child back to the Infirmary.

There was no vasomotor mottling of the skin.

The sensory functions were perfectly normal. There was no affection of the bladder or rectum.

The heart and other organs were all healthy. The body was covered with a considerable quantity of subcutaneous fat.

Family History.—The parents were healthy, robust country people; no case of a similar kind had ever, so far as they knew, occurred amongst any of their relatives.

Diagnosis.—The case appeared to me to be very similar to the case described by Drs. Thomson and Bruce (see page 264). Accordingly, after examining the patient, I sent him on to Dr. John Thomson, but without giving any indication as to what I thought of the nature of the condition. Dr. Thomson wrote me as follows:—"It is a most interesting case. I never saw one exactly like it, but I think it has more points of resemblance to that of the little girl which Bruce and I are publishing in the Hospital Reports than to anything else that I have seen. Many of your patient's symptoms are just the same, but his back and arms are more severely affected than in our case. If I am not mistaken, the lower part of the pectoralis major is deficient or wanting; in our case it was very distinctly present. I should be inclined to diagnose the case as one of progressive muscular dystrophy, whether with or without a spinal lesion I do not know."

Duration and course.—The patient remained much *in statu*

quo until February 1894, when he died after four days' illness from acute pneumonia. Dr. MacAllister, who had done his best to persuade the parents to bring him to Edinburgh, informed me that the trunk and leg muscles gradually became more atrophied and flabby, but that there seemed to be some improvement in the condition of the arms; for a few months previous to death, the patient had been able to use his arms and hands a little more than he had been able to do previously.

A post-mortem examination was not allowed.

THE PERONEAL TYPE OF PROGRESSIVE MUSCULAR ATROPHY.

Dr. Tooth has applied this term to certain cases of muscular atrophy in which the muscular atrophy begins in the small muscles of the leg. Charcot, Marie, Hoffmann and others have described similar cases. Hoffman has proposed the term '*progressive neurotic atrophy*'; and the disease is sometimes termed '*the Charcot-Marie type of progressive muscular atrophy*.'

The disease is very rare; one case only has come under my own observation. Its exact pathology is not as yet definitely understood; but it undoubtedly seems to be quite distinct from the cases of myopathic atrophy which I have just described. The clinical features of the disease seem to show that the lesion is situated either in the peripheral nerves or in the spinal cord. The atrophy is consequently either neuropathic or myelopathic.

As a rule, the disease begins in later childhood, youth, or early adult life. Males are more frequently affected than females; but the tendency to affect males in preference to females is less marked than in pseudo-hypertrophic paralysis.

In a considerable proportion of cases, the disease seems to be hereditary, or to attack several members of the same family; isolated cases are probably more frequent than isolated cases of the myopathic forms of atrophy. In some cases the disease has followed an attack of measles.

The course of the affection is very chronic. The atrophy commences in the muscles of the foot or leg. The extensor hallucis pollicis is usually first affected; then the extensors of the toes, the peronei and the small muscles of the foot are

involved. It is not improbable, however, that in many cases the small muscles of the foot are affected at the very commencement, for weakness and atrophy in these muscles are much less easily detected than in the muscles of the leg, and are, therefore, apt to escape attention.

The atrophy is developed slowly and gradually and is progressive. After a time, the calf muscles become affected, and a characteristic deformity—talipes varus—which is due to weakness of the peronei muscles is developed.

After the atrophy has existed for some time (it may be three or four years or even longer) in the lower extremities, the muscles of the upper extremity become involved. The small muscles of the hand (thenar and hypothenar muscles) and the muscles of the forearm are first affected; and the bird-claw appearance of the hand which is so characteristic of the Aran-Duchenne type of progressive muscular atrophy is produced.

The muscles of the thigh, upper arm, pelvic and shoulder girdles, back, trunk and neck, may ultimately become involved.

The atrophy is usually but not always symmetrical. In the first case, for example, reported by Tooth in his memoir on the subject, the muscles of the right leg were markedly atrophied, while those of the left were scarcely affected. There is never any muscular hypertrophy or pseudo-hypertrophy.

Fibrillary or fascicular muscular twitchings frequently occur, and the reaction of degeneration in its imperfect form is usually developed. Myalgic pains occasionally occur, and various derangements of sensation are also sometimes observed.

It is obvious from these characters that the type of disease is peculiar. It resembles the Aran-Duchenne type in the facts that the muscles of the distal end of the affected extremities are first involved, and that fibrillary twitchings and the reaction of degeneration are developed. But it differs from the Aran-Duchenne type inasmuch as the atrophy begins in the muscles of the foot and leg, that the disease usually commences in childhood, youth, or early adult life, and that in many cases it presents the hereditary and family tendencies, which are so characteristic of the myopathic forms of muscular atrophy.

As yet, very few cases have been examined post mortem. In two at least, changes in the peripheral nerves, without any corresponding lesions in the anterior cornua of grey matter, were

present. But it is perhaps, as yet, premature to conclude that the lesion is neuropathic. As Sachs has pointed out, the clinical features of the disease closely correspond to those of progressive muscular atrophy of the Aran-Duchenne type. Sachs indeed proposes to term the disease 'the leg type of progressive muscular atrophy.' Further pathological observations are required in order to settle this disputed point.

Diagnosis.—When well advanced, the disease is easily recognised. The bird-claw condition of the hands, when combined with talipes varus and atrophy of the muscles of the leg and foot, is highly suggestive. If, in a case of this kind, it is ascertained that the atrophy commenced slowly and gradually in the muscles of the foot and leg and subsequently extended after a considerable interval to the muscles of the hands and forearms, the diagnosis of the peroneal type of progressive muscular atrophy may confidently be made.

Prognosis.—The course of the disease is slow and chronic, but, so far as our present knowledge enables us to judge, usually progressive.

Treatment.—The same treatment which has been recommended for the Aran-Duchenne type of progressive muscular atrophy should be employed. An elastic bandage and stiff boot are advisable in order to counteract the foot deformity.

LECTURE XVI

LOCOMOTOR ATAXIA

TO-DAY, Gentlemen, I propose to commence the consideration of locomotor ataxia, one of the most important subjects which we shall have to consider in this Course, for the disease is very common. The symptoms which it presents are very numerous and diverse, the suffering which it entails is in many cases very great, and the management of cases of locomotor ataxia often imposes a heavy tax upon the therapeutic resources of the practitioner.

Synonyms. — *Tabes dorsalis*, *Sclerosis of the posterior columns of the spinal cord*, *Progressive locomotor ataxia* and *Neuro-spinal tabes* are some of the synonyms which have been applied to the disease.

Morbid Anatomy and Pathology.—Let us first look at the morbid anatomy and pathology. The disease is due to a lesion of the afferent or ingoing half of the nerve apparatus. The lesion is not confined to the spinal cord, for the sensory fibres of the peripheral nerves are in many cases involved. Hence the term *neuro-spinal tabes* (a wasting of the nerves and spinal cord) which has recently been given to the disease.

Again, it is important to remember that in many cases of locomotor ataxia temporary paralyses of the ocular muscles are developed; and that in other cases loss of sight from atrophy of the optic nerve or some derangement of hearing from implication of the auditory nerve or its terminations occur. In short, in cases of locomotor ataxia, the lesions are much more extensively distributed through the nervous system than was at one time supposed.

Further information is required as to the condition of the

sensory nerves in the muscles in cases of locomotor ataxia and as to the relative importance (so far as the production of the symptoms is concerned) of the lesion in the spinal cord and in the sensory nerves of the muscles. But so far as our present knowledge enables us to judge, the essential and fundamental pathological substratum of locomotor ataxia is a sclerosis of the posterior columns of the spinal cord. It must, however, be stated that many of the symptoms which characterise locomotor ataxia may be developed entirely as the result of a lesion of the peripheral nerves; in other words, in cases in which the posterior columns of the cord are healthy. To these cases the terms *pseudo-tabes* and *neuro-tabes périphérique* are sometimes applied.

Naked-eye appearances of the spinal cord and nerve roots.—

In the advanced stages of the disease, the lower part of the cord usually looks somewhat compressed and flattened in its antero-posterior diameter, and the posterior columns appear to be less plump than normal; the membranes over the posterior surface of the cord are in some cases distinctly thickened; but the most conspicuous change is usually the atrophied condition of the posterior nerve roots in the lumbar and lower dorsal regions. The contrast between the normal anterior root-bundles and the grey, shrivelled and atrophied posterior roots is usually very marked. This is the exact reverse, you will observe, of the condition which characterises progressive muscular atrophy. In that disease, the most striking naked-eye alteration is the atrophied and wasted condition of the anterior nerve roots; in that disease, the posterior nerve roots are plump and healthy. Another point of distinction is this, that in progressive muscular atrophy the anterior nerve roots attached to the cervical enlargement are chiefly involved; whereas in locomotor ataxia the posterior nerve roots attached to the lumbar enlargement are chiefly affected.

On cutting across the lumbar region of a cord affected with locomotor ataxia, the lesion in the posterior columns is more apparent; the posterior columns are seen to have a grey, translucent appearance, and their consistency is increased. After hardening with suitable reagents, the diseased condition of the posterior columns becomes still more conspicuous.

Microscopic appearances.—On microscopical examination of properly hardened sections, the sclerosis in the posterior columns

is very apparent, for the sclerosed portions stain vividly with carmine and only very slightly with osmic acid or by Weigert's method; whereas the posterior columns of the healthy cord are only slightly stained with carmine, while they are deeply stained with osmic acid and by Weigert's method.

In old-standing cases—for it is seldom that the opportunity occurs of examining the lesion in the early stages—the whole thickness of the posterior columns (both the postero-internal and the postero-external columns) is more or less completely destroyed in the lumbar and lower dorsal regions.

In the upper dorsal and cervical regions, the lesion is usually limited to the postero-internal columns (the columns of Goll) and presents the usual characters of a secondary ascending degeneration.

These appearances are well seen in figs. 91, 92 and 93.

In exceptional cases (see fig. 95) the postero-external columns in the cervical region are sclerosed, but the lesion of the postero-external columns is rarely so extensive in the cervical as in the lumbar region.

Under a high power, we find that the nerve fibres in the affected portions of the cord have more or less completely disappeared; they are replaced by a delicate fibrillated connective tissue. The connective tissue trabeculae are increased in size, the Deiters' cells enlarged, and the walls of the blood-vessels in the affected parts of the cord are usually more or less thickened. Corpora amylacea are in some cases scattered in great abundance through the degenerated tissue (see fig. 96).¹

In some cases, the pia mater over the posterior columns, sometimes round the whole surface of the cord, is thickened, evidently as the result of a chronic meningitis.

In advanced stages of the disease, the grey matter of the posterior horn is usually implicated by the sclerotic process. In some cases the nerve cells in the posterior horn are atrophied or vacuolated. The vesicular column of Clarke and the adjacent grey matter, which lies between the anterior and posterior cornua, are also sometimes involved.

¹ These corpora amylacea, which stain deeply with carmine but which are unstained by osmic acid, are not peculiar to locomotor ataxia; they occur in many other conditions in which nerve tubes are undergoing degenerative or inflammatory destruction.

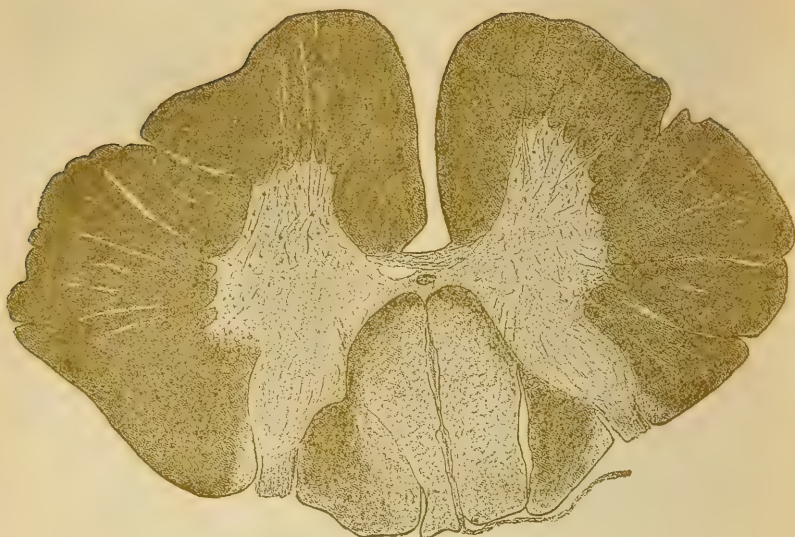


FIG. 91.—*Transverse section through the cervical enlargement in a case of locomotor ataxia. Stained with osmic acid, mounted in Farrant's solution, and magnified about 10 diameters.*

The lesion is confined (or almost so) to the postero-internal columns; and presents the usual features of a secondary ascending degeneration.



FIG. 92.—*Transverse section through the dorsal region of the cord in a case of locomotor ataxia. Stained with osmic acid, mounted in Farrant's solution, and magnified about 10 diameters.*

The lesion involves the whole of the posterior columns, which are seen to be very faintly stained by the acid.

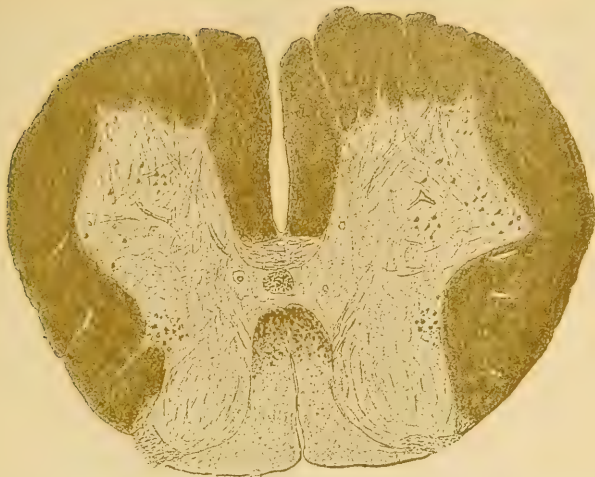


FIG. 93.—*Transverse section through the lumbar region of the cord in a case of locomotor ataxia. Stained with osmic acid, mounted in Farrant's solution, and magnified about 10 diameters.*

The greater part of the posterior columns is invaded by the lesion, and is, therefore, unstained by the acid. The part of the posterior columns adjacent to the posterior commissure is still healthy.

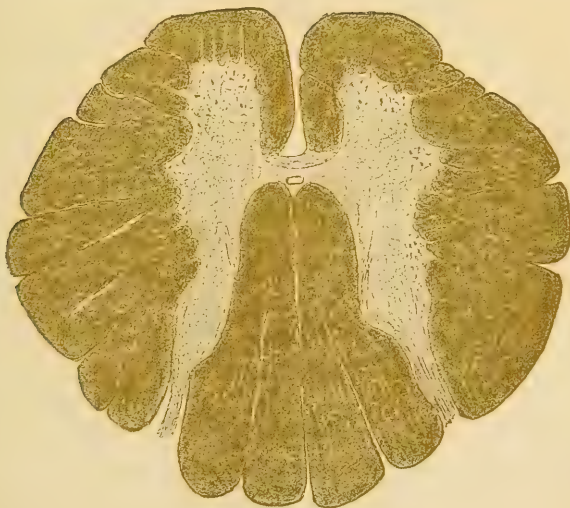


FIG. 94.—*Transverse section through the dorsal region of a healthy spinal cord. Stained with osmic acid, mounted in Farrant's solution, and magnified about 10 diameters.*

All the white columns are deeply stained by the acid. The grey matter is only slightly affected by it.

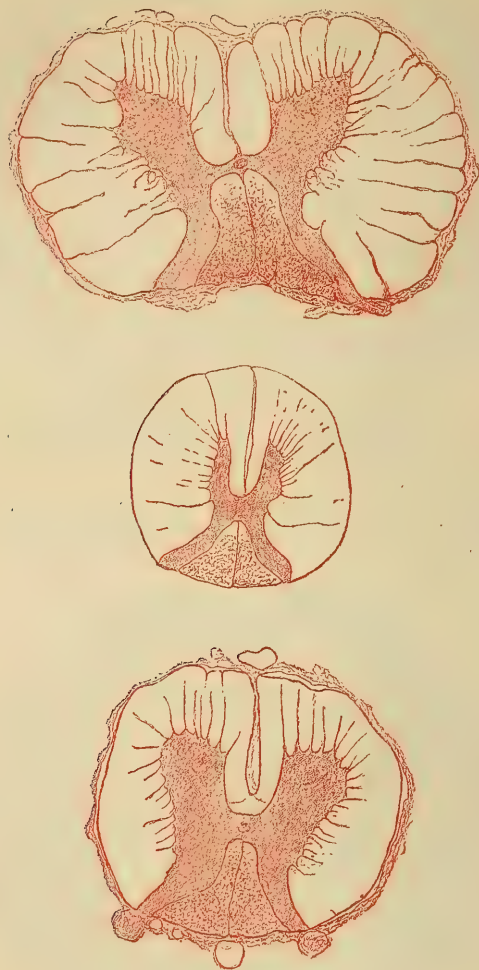


FIG. 95.—*Transverse sections through the cervical, dorsal and lumbar regions of the spinal cord in a case of locomotor ataxia.*

The postero-external columns and Lissauer's tracts are sclerosed in the cervical as well as in the dorsal and lumbar regions. The ataxia and lightning pains affected the arms as well as the legs. The pia mater over the posterior columns is markedly thickened and inflamed; and thick bands of connective tissue pass from the thickened pia into the posterior columns.

The atrophy of the posterior roots, which when slight is more apparent on microscopical than on naked-eye examination, stops short at the posterior root-ganglia. That is a significant fact. It suggests that the atrophy of the posterior roots, and perhaps also the sclerotic lesion in the cord, is of the nature of a secondary ascending degeneration.

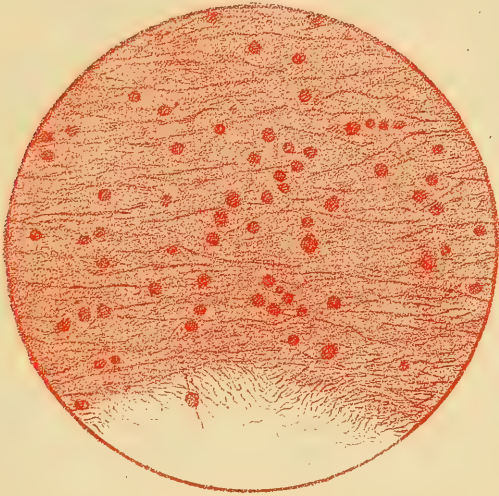


FIG. 96.—*Longitudinal section through the posterior column of the cord in locomotor ataxia. Stained with carmine, mounted in dammar, and magnified about 250 diameters.*

The nerve tubes have almost entirely disappeared. Their place is taken by a delicate connective tissue. Numerous corpora amylacea are scattered through the section. These bodies are deeply stained by carmine and logwood, faintly by osmic acid.

We have seen that the fibres of the pyramidal tract are nourished by the large pyramidal cells in the motor area of the cortex of the brain, and that the motor fibres of the peripheral nerves are nourished by the multipolar nerve cells of the anterior cornu of the spinal cord. I have now to add that the fibres of the posterior columns of the spinal cord are nourished by the nerve cells in the posterior root-ganglia. Destruction of the posterior root-ganglia or of the posterior nerve roots between the ganglia and the spinal cord produces an ascending degeneration which passes through the postero-external column of the segment

of the cord to which the affected nerve root is attached and which extends from the seat of the lesion right up to the top of the cord in the postero-internal column.

And this leads me to say that opinions differ as to the exact part of the ingoing nerve apparatus which is first affected. Some authorities believe that the lesion commences in the postero-external columns of the spinal cord; others in the posterior roots or the posterior root-ganglia; others again in the peripheral nerves.

In typical cases the cord lesion seems to be a chronic inflammation, which, commencing in the postero-external columns in the lumbar region, gradually invades and ultimately destroys the whole of the posterior columns in the lumbar and lower dorsal regions; and which is associated with a secondary ascending degeneration of the postero-internal columns in the upper part of the cord (the segments of the cord which are situated above the lesion). According to this view, the thickening (sclerosis) of the connective tissue and blood-vessels in the affected parts of the cord is secondary.

Another view supposes that the lesion commences in the interstitial connective tissue, and that as the sclerosis advances, the nerve tubes are, as it were, strangled and destroyed.

Quite recently Dr. Nageotte (reported in the *Medical Week*, Nov. 17th, 1894; p. 561) has advanced the view that the primary lesion is a very intense perineuritis, which occupies exactly that position of the pair of spinal nerves which is situated between the ganglion and the entrance of the roots into the arachnoid cavity. This perineuritis which is at first embryonic, but changes to fibroid at an advanced stage, is, he thinks, primary and leads secondarily to degeneration of the fibres of the posterior roots.

A fourth view supposes that the lesion commences as a meningitis on the surface of the cord and that the inflammatory process passes into the cord along the connective tissue septa and vessels.

While a fifth view is that which I have already indicated, namely, that the cord lesion is a secondary ascending degeneration, due to a lesion in the posterior root-ganglia or in the peripheral nerve-endings.

It is possible that all of these views may be, in some degree

correct; in other words, that different conditions may in different cases contribute to the production of the cord lesion; but the first view, which supposes that the disease commences as a parenchymatous inflammation of the nerve fibres in the postero-external columns of the spinal cord, seems to me, in the present position of our knowledge, the most satisfactory.

In some cases which have been examined post mortem, the posterior root-ganglia and posterior roots have been found free from disease.

According to Pierret, the lesion commences in a particular

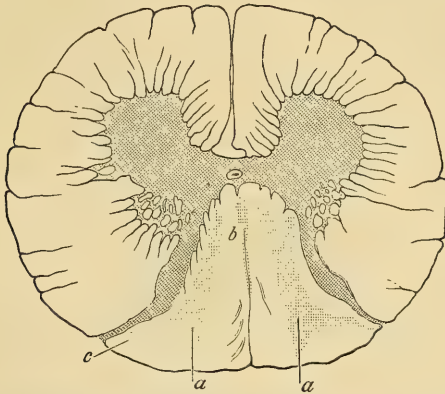


FIG. 97.—Transverse section through the spinal cord in an early stage of locomotor ataxia, showing 'sclerose des bandelettes externes.'—(After Pierret).

The letters *a* point to *les bandelettes externes*. The letter *b* points to a thin line of degeneration situated a short distance behind the grey commissure on each side of the posterior median fissure. The letter *c* points to Lissauer's column.

part of the postero-external column—*les bandelettes externes* (see fig. 97). I must not forget to add that Lissauer has shown that the fine bundles of nerve fibres which lie on each side of the posterior roots, and which are known by the term of Lissauer's tract or column of fibres (see fig. 98), are affected (sclerosed) even in the early stages of the disease.

Before leaving the morbid anatomy, let me say that the direct cerebellar tracts and the ascending lateral tracts of Gowers are in some cases also sclerosed. Both of these tracts conduct ingoing or upward-going impressions. The fibres of the direct cerebellar tract are nourished by (have their trophic centres in)

the nerve cells of Clarke's vesicular column. The sclerosis of the direct cerebellar tracts seems, like the sclerosis of the columns of Goll, to be a secondary ascending degeneration. You will readily understand, therefore, that the sclerosis of the direct cerebellar tracts will only be present in those cases in which the vesicular columns of Clarke are implicated by the lesion.

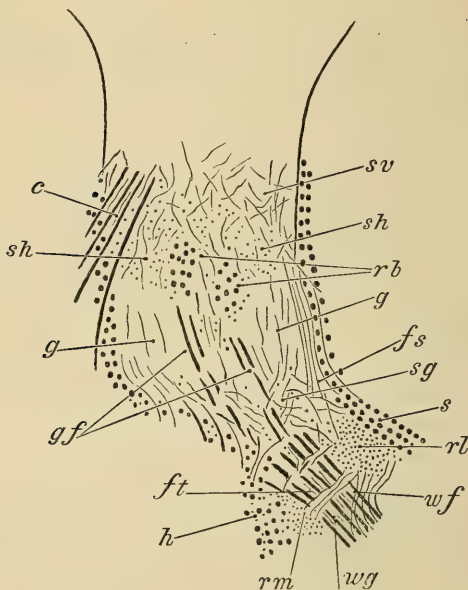


FIG. 98.—*The posterior root and posterior horn of grey matter with the adjacent parts of the lateral and posterior columns.*—(After Lissauer.)

h. Commencement of the posterior column; *s.* commencement of the lateral column; *wg.* posterior root at its point of entrance into the cord; *wf.* fine radicular fibres; *rl.* lateral, and *rm.* median part of the marginal zone (Lissauer's column); *ft.* fasciculus of transverse fibres traversing the posterior root at its point of entrance into the cord; *sg.* spongy zone of the gelatinous substance; *fs.* fibres which seem to connect the spongy substance with the lateral column; *g.* typical gelatinous substance; *gf.* large fibres which pass directly and in bundles through the gelatinous substance; *sh.* posterior segment of the gelatinous substance; *rb.* longitudinal fibres of large size; *sv.* anterior segment of the spongy substance; *e.* fibres passing from the posterior column into the posterior horn of grey matter.

Further, in the later stages of the disease, the lesion may extend to the crossed pyramidal tract in the lateral column or

to the anterior cornu of grey matter. But the implication of these parts of the cord is, as it were, accidental—an associated complication which is in no way a characteristic and essential feature of the disease.

Up to this point I have been speaking of the morbid changes in the spinal cord. I must now ask you to note that the lesion seems rarely, if ever, limited to the spinal cord. In many cases, the peripheral sensory nerves are also affected by what seems to be a chronic inflammatory process—a parenchymatous inflammation—which is probably identical with that which affects the nerve fibres of the postero-external columns of the cord.

The intensity of this peripheral neuritis varies in different cases. In some cases, the peripheral nerve trunks are affected; but in many cases the terminal fibres of the sensory nerves, more especially perhaps the fine sensory nerves of the muscles, seem to be alone or chiefly implicated.

The implication of the fine sensory nerves of the muscles of the lower extremities is of great importance from a clinical point of view; for the integrity of the muscular sense, and therefore of co-ordination, without doubt largely depend upon the condition of the fine sensory nerves of the muscles.

In the present position of our knowledge, it is hardly possible to make a definite statement as regards the frequency with which the peripheral nerves are involved. It is only of recent years that attention has been directed to the condition of the peripheral nerves. The detection of minute changes in the fine sensory nerve terminations is a very difficult matter. Until a larger number of cases have been examined by competent observers, thoroughly skilled in modern methods of research, we must be content to remain in doubt as to the frequency with which the sensory nerve terminations in the muscles are affected, and as to the part which the lesion of these nerves plays in the production of the clinical phenomena of the disease.

In a certain proportion of cases of locomotor ataxia, the optic nerve undergoes a condition of grey atrophy; this is of great importance from a clinical point of view. The roots of the vagi and of the sensory roots of the fifth nerve are sometimes sclerosed and atrophied. Indeed, it is safe to say that every sensory or ingoing nerve tract may be affected.

Pathological Physiology.—Let us turn now to the pathological physiology. We have seen that the lesion is confined to the afferent side of the nerve apparatus, to nerves and tracts which carry impulses inwards and upwards. The efferent tracts and nerves, which conduct impulses downwards and outwards, are unaffected. I am speaking of course of typical and uncomplicated cases of the disease. So far as our present knowledge enables us to judge, the sclerosis of the posterior columns of the cord is the fundamental and essential feature of the disease; but it is not improbable that the lesion of the peripheral nerves, especially of the fine sensory nerves of the muscles, plays a more important part in the production of the symptoms than has been hitherto supposed.

Now, since the lesion is confined to the afferent (sensory) side of the nerve apparatus, the chief symptoms will necessarily consist of disturbances of sensation, of reflex action and of co-ordination. Motor symptoms are conspicuous by their absence. There is no paralysis, no muscular weakness, no muscular atrophy.

There is one apparent exception to this statement, for temporary paralysis of one or other of the ocular muscles is common in the early stages of the disease. The exact cause of these passing paralyses of the ocular muscles is not clear.

Occasionally, too, paralysis of the limbs, which may be hemiplegic, paraplegic or monoplegic in distribution, occurs. The causes of these paralytic complications is not always apparent. In some cases, the paralysis is temporary and apparently analogous to the temporary paralysis of the ocular muscles to which I have just referred. In other cases in which the paralysis is more persistent, or it may even be permanent, inflammation of the peripheral motor nerves, extension of the cord lesion to the lateral columns or anterior cornua, and associated syphilitic lesions of the vessels of the brain or extravasations of blood into the brain tissue seem to be the chief causes of the paralysis.

You must remember that reflex action and co-ordination are readily deranged by lesions entirely confined to the afferent side of the nerve apparatus. Loss of the knee-jerks, lightning pains, loss of the pupil reflex to light, inco-ordination, impairment or loss of the muscular sense, partial anæsthesia and hyperæsthesia,

and derangement of the vesical, rectal and sexual reflexes are the most conspicuous symptoms of the disease. All of these symptoms are easily accounted for by the lesion in the posterior columns of the cord and in the sensory peripheral nerves.

Further, the lesion for the most part is confined to the lower part of the spinal cord¹ and to the sensory peripheral nerves connected with the lumbar and sacral regions of the cord. Hence, the symptoms are, for the most part, confined to the lower extremities and the pelvic viscera.

The symptoms enumerated above do not by any means include all of the symptoms which may be present in cases of locomotor ataxia. They are merely the more common and outstanding symptoms.

The exact manner in which the various symptoms which characterise the disease are produced will be more appropriately considered in connection with the clinical history. In describing the clinical history I will refer in more detail to the pathological physiology, and will endeavour to explain to you the manner in which the individual symptoms are produced.

Etiology.—Locomotor ataxia is essentially a disease of the adult male; females are, comparatively speaking, rarely affected. The disease usually commences between the ages of thirty and forty-five; it very rarely develops before the age of twenty, and it is almost unknown before the age of puberty. A few undoubted cases have, it is true, been met with in younger persons, and it is interesting to note that in these cases the patients have always, it is said, been the subjects of inherited syphilis.²

And this leads me to say that syphilis is *the* most important etiological factor in the production of locomotor ataxia. A very large proportion of the patients who are affected with locomotor ataxia have previously suffered from constitutional syphilis. Unfortunately, as we shall afterwards see, the fact that a patient has had syphilis does not in any material way improve the prognosis or help our treatment; for locomotor ataxia, like

¹ The sclerosis of Goll's column is merely a secondary ascending degeneration of fibres which are directly continuous with the affected fibres of the postero-external columns.

² Friedreich's ataxia, a disease which occurs in children and which is not due to inherited syphilis, must not be confounded with locomotor ataxia.

general paralysis of the insane, although undoubtedly a result of syphilis, is not a *direct* result of the disease; it is not directly influenced and cured by iodide of potassium and mercury.

It is difficult to state the exact frequency with which locomotor ataxia is preceded by syphilis; for, in some cases in which no definite and distinct history of syphilis can be elicited, it is certain that the patients have suffered from the disease. But it is probably not far from the truth to say that in at least eighty-five per cent. of the cases of locomotor ataxia the patients have had syphilis. Some authorities go further than this. Dr. Drummond, for example, has advocated the view that every case of locomotor ataxia is the result of previous syphilis. But with this opinion I cannot agree. I of course admit the difficulty that there often is in eliciting a history of former syphilis. I allow that in a certain proportion of the cases of locomotor ataxia in which a syphilitic history cannot be elicited the patients have had syphilis. But this is a very different thing from saying that the patient has had syphilis in every case of locomotor ataxia in which syphilis is denied, forgotten or not noticed. More than one case has come under my own notice in which I feel certain (so far as one can be certain about any fact which has not come under one's own personal observation) that there were no syphilitic antecedents. I see no reason to conclude that, because 80 or even 90 per cent. of patients affected with locomotor ataxia have had syphilis, the remaining 20 or even 10 per cent. *must* (logically speaking) have had syphilis. We know that peripheral neuritis may be due to a variety of toxic irritants; and it is reasonable to suppose that the parenchymatous neuritis of the fibres of the postero-external columns of the cord, which seems to be the essential pathological substratum of locomotor ataxia, may be produced by more than one irritant.

The exact manner in which previous syphilis leads to the production of locomotor ataxia is not definitely known. The most probable view is that the organism of syphilis generates a chemical poison, a toxine, which has a special irritative action upon the nerve fibres of the posterior columns of the cord and of the sensory terminations of the peripheral nerves, just as one of the toxines which is produced by the diphtheria bacillus undoubtedly causes an inflammation of the peripheral nerves.

The weak point in this theory seems to me to be the fact that the fibres of the postero-external columns in the cervical region usually escape. But this difficulty is removed if we suppose that more factors than one come into play in the production of the disease.

Further, we know that the toxine of the diphtheria bacillus has a very selective action. I do not refer so much to the throat paralysis (for the inflammation of the nerves of the palate may perhaps be explained by the local concentration of the poison), but to the paralysis of the ciliary muscle.

It seems to me not improbable either (1) that the toxine produced by the syphilitic organism may exert a depressing or irritative effect upon the ingoing or afferent nerve apparatus as a whole, which predisposes it to be injuriously affected by other irritants and depressing causes (such as sexual excess) which have a selective action upon the part of the ingoing apparatus which is situated in the lower part of the spinal cord; or *vice versâ*, (2) that these depressing causes (sexual excesses, etc.), which act on the lower part of the spinal cord, may predispose it (the lower part of the cord) to be affected by the toxine produced by the syphilitic organism.

Further, in trying to explain why the lower parts of the cord should be so much more frequently affected than the upper parts, the comparative difficulty with which the lower part of the spinal cord is supplied with blood must be borne in mind. (Dr. Moxon's observations on this point are referred to on page 21.)

In one or other of these ways the limitation of the lesion to special parts of the ingoing apparatus and to special parts of the spinal cord may perhaps be most satisfactorily explained.

The fact that males suffer from locomotor ataxia so much more frequently than females is no doubt largely due to the circumstance that they contract syphilis much more frequently than females. But this is not perhaps the sole explanation. Almost every chronic disease of the spinal cord, which is acquired in adult life, is more frequent in the male than in the female. We have already noted this fact in connection with progressive muscular atrophy and primary spastic paraplegia. It would appear that the spinal cord of the male is more exposed to conditions which favour the development of inflammatory, sclerotic and degenerative processes than the spinal

cord of the female. Leaving syphilis out of account—and speaking generally—males are more exposed than females to neuromuscular strain (mental strain, anxiety, prolonged worry, general wear and tear, etc.), cold, alcoholic and sexual excesses, peripheral irritations of all kinds and traumatic injuries. Further, we know that some other sclerotic lesions, such, for example, as cirrhosis of the liver, kidney and heart, are more common in men than in women. No doubt the facts that men drink more alcohol and more frequently suffer from syphilis go far to explain these differences. The fact that men suffer much more frequently from gout and the degenerative lesions produced by gout is probably not unimportant; for, as every one knows, the circulation in the blood of the gouty poison is the chief cause of cirrhosis of the kidney.

All of these circumstances, and especially perhaps the fact that sexual excess acts much more injuriously upon the male than upon the female, and upon the lower than upon the upper part of the spinal mechanism, have probably some bearing upon the etiology of locomotor ataxia and afford an explanation of the greater liability of the male.

Locomotor ataxia is more common in the well-to-do and professional classes than in the lower orders of society. It is much more common in people who live in towns than in people who live in the country, probably because syphilis is more common in towns than in country districts.

The disease seems to occur in all climates and amongst all peoples. The Jewish race, who very rarely suffer from syphilis, are very seldom affected.

I repeat that, while syphilis is the most important factor in the production of locomotor ataxia, other contributory causes must be taken into account. We see the same thing in the case of some other lesions which are more definitely and distinctly syphilitic than the lesion of locomotor ataxia. A tertiary gumma on the surface of the brain often owes its immediate development to a blow on the head or to mental strain—to something in short, which disturbs the circulation and the vascularity of the membranes of the brain. Again, arterial strain is apt to produce arterial disease and aneurisms in persons who are the subjects of syphilis. It is quite reasonable, therefore, to suppose that the sclerotic lesions of general paralysis of the insane and of loco-

motor ataxia are in some cases excited by brain strain or cord strain, and by conditions (such as sexual excesses, blows on the head and back, exposure to cold and wet, etc.) which disturb the vascularity of the brain or the spinal cord, or which seriously derange the functional activity and the metabolism of the nerve elements in which the sclerotic lesion is subsequently developed.

These considerations are of great importance, for they have a direct practical bearing upon the prevention of locomotor ataxia and general paralysis of the insane in persons who have previously suffered from constitutional syphilis.

Locomotor ataxia is very rarely indeed directly inherited. In a certain proportion of cases, patients affected with locomotor ataxia come of a nervous (neurotic) stock, and some authorities have laid great stress upon this indirect neurotic inheritance as a predisposing cause of the disease; it probably to some extent accounts for the fact that one man who has contracted syphilis will subsequently become affected with locomotor ataxia, while another will escape. A hereditary tendency to diabetes perhaps also predisposes to the production of locomotor ataxia. Some French authorities attach considerable importance to this connection.

Let us now pass to the clinical history.

Clinical History.—The clinical picture which different cases of locomotor ataxia present is very different, for the symptoms are multifarious and the manner in which they are grouped is subject to great variations in different cases. Further, the mode of onset and the rapidity with which the symptoms develop and the disease progresses are very variable. Nevertheless, most cases conform to certain well-recognised types.

For the purposes of description and arrangement, we are in the habit of dividing the disease into three stages:—(1) the stage of invasion; (2) the stage of full development; and (3) the stage of complications and extension. These stages are not always sharply defined; in many cases they run insensibly one into another.

Another subdivision, which is of the greatest practical importance, for it represents not merely two stages of the disease, but two distinct clinical types which are true to nature, divides

the disease into two stages, viz., (1) a pre-ataxic stage, and (2) an ataxic stage.

Further, we may divide the ataxic stage into two sub-stages, viz., (a) an ataxic stage in which the patient is able to walk, and (b) an ataxic stage in which he is unable to walk.

In most cases of locomotor ataxia—the ordinary typical cases—the disease commences with a pre-ataxic stage, which is subsequently followed by an ataxic stage.

But all cases are not typical.

In some cases, the symptoms of the pre-ataxic and ataxic stages are developed simultaneously, or at all events the ataxic stage is not preceded, so far as our observation and information enable us to judge, by a definite pre-ataxic stage. In these cases, the ataxia is an early symptom, though it is perhaps not the first symptom, for it is probably preceded in most (if not in all) cases by loss of the knee-jerks, and (it probably would be found to be preceded in the great majority of cases if an absolute accurate history were forthcoming) by lightning pains.

In other cases, the duration of the pre-ataxic stage is unusually long; in fact, in rare cases the ataxic stage seems to be never reached. The cases included in this group form a well-marked clinical type of the disease—the *pre-ataxic form* of tabes dorsalis.

We may say then that there are three leading clinical types of locomotor ataxia, viz. :—

(1) The ordinary typical cases in which a well-marked pre-ataxic stage is followed after an interval of some, it may be many, months by a typical ataxic stage.

(2) Cases in which the pre-ataxic stage is unusually long, and in some of which the ataxic stage is perhaps never developed (cases of pre-ataxic tabes).

(3) Cases in which the pre-ataxic stage is unusually short or (apparently) altogether absent, and in which the ataxic symptoms appear to be developed quite at the commencement of the disease.

The symptoms of the pre-ataxic stage.—The chief symptoms which characterise the pre-ataxic stage or the stage of invasion are :—Loss of the knee-jerks, lightning pains, and loss of the pupil reflex to light. Some disturbance of the vesical, rectal and sexual reflexes (paresis of the bladder, constipation, diminished

or increased sexual desire and sexual power), temporary paralysis of some of the external muscles of the eyeball (such as ptosis) and some derangement of the tactile sensibility (hyperæsthesia and anæsthesia), are often present during this stage of the disease.

Optic atrophy, gastric and other forms of visceral crises, and trophic lesions in the bones and joints, etc., which are occasional, but, comparatively speaking, rare symptoms of the disease, are, when they do occur, almost always developed during this, the early, pre-ataxic, stage of the disease.

The symptoms of the ataxic stage.—The chief symptoms which are characteristic of the ataxic or fully developed stage of the disease are:—Difficulty in walking, inco-ordination, impairment or loss of the muscular sense, and inability to balance the body in the erect position with the eyes closed (Romberg's symptom).

During the second or ataxic stage of the disease, the symptoms which are characteristic of the pre-ataxic or first stage of the disease not only continue but are apt to increase in severity. The derangements of the bladder, rectum and sexual apparatus, and the anæsthesia almost always become aggravated as the second stage of the disease advances.

Some of the symptoms which have been enumerated as characteristic of the pre-ataxic stage of the disease may only be developed when the ataxic or second stage of the disease is reached. And it must be remembered that all of the symptoms which have been enumerated above are not present in every case of locomotor ataxia. The lightning pains and the Argyll Robertson condition of the pupil, for example—two of the most constant and important symptoms of the pre-ataxic stage—are in some cases never developed; while some of the rarer symptoms, such as the gastric crises, optic atrophy and joint lesions, are only developed in a small proportion of cases.

From this statement, it will be apparent that the second or ataxic stage of the disease is characterised by the presence of:—(1) the (permanent or persisting) symptoms of the first or pre-ataxic stage, which happen to be present in the individual case under consideration; *plus* (2) the characteristic symptoms of the second or ataxic stage, viz., difficulty in walking, ataxia, impairment of the muscular sense, Romberg's symptom, etc.

Some of the symptoms of the pre-ataxic stage, such as the loss of the knee-jerks, loss of the pupil reflex to light, optic atrophy and Charcot's joint lesion, may be termed permanent or persisting symptoms ; for (with very rare exceptions) they continue and persist once they are developed.

Others, such as the temporary paralysis of the ocular muscles and the herpetic or other eruptions on the skin, may be termed temporary or fleeting symptoms, for they are of brief duration, though in some cases they recur from time to time.

Others, such as the lightning pains and visceral crises, occupy a mid-position. The paroxysms are temporary, but they almost always continue to recur ; they not unfrequently disappear, however, or diminish in severity as the disease progresses ; the latter statement applies especially to the visceral crises. In one sense, therefore, they may be said to persist.

LECTURE XVII

LOCOMOTOR ATAXIA (*Continued*)

IN the last lecture, Gentlemen, I described in outline the more important clinical features of locomotor ataxia. Let us now consider the individual symptoms in detail.

Difficulty in walking, ataxia, inco-ordination.—Let us suppose that we are dealing with a typical and fully developed case of the disease. The most striking symptom in such a case is difficulty in walking and a peculiar and characteristic gait. The difficulty in walking is not due to loss of muscular power, but to defective co-ordination. We have seen that the lesion is confined to the ingoing (afferent) side of the nerve apparatus. The outgoing or motor side is unaffected. There is consequently no paralysis, no muscular atrophy. When the ataxic stage is fully developed and the difficulty in walking very marked, the muscles of the legs may be firm and well nourished and the gross muscular force very great.

I repeat that the difficulty in walking is due to inco-ordination and that the inco-ordination seems to be the result of the lesion in the ingoing nerve apparatus—the lesion in:—(1) the postero-external columns of the spinal cord; and (2) the fine peripheral terminations of the sensory nerves, especially of the sensory nerves of the muscles.

In the earlier stages, and in fact in most cases throughout the whole course of the disease, the lesion is, in the great majority of cases, confined, or for practical purposes confined, to the lower end of the spinal cord, for the sclerotic lesion in the columns of Goll which extends up to the top of the cord is merely a secondary ascending degeneration, which affects those fibres of the postero-internal column (the column of Goll) which are the upward continuations of the fibres of the postero-external column *in the lumbar and lower dorsal regions*.

The inco-ordination is consequently in the earlier stages, and in some cases throughout the whole course of the disease, limited to the lower extremities.

In other cases, after first affecting the lower extremities, the inco-ordination ultimately involves, though seldom in a marked degree, the upper limbs. In cases of this kind, the lesion pursues an ascending course. It is only in very rare cases that the ataxia is first developed in the upper limbs and subsequently extends to the lower extremities. In other words, it is quite exceptional for the lesion to pursue a descending course.

The explanation of some of these facts will be more apparent after we have considered the exact manner in which the inco-ordination is produced.

The inco-ordination is in most cases developed insidiously and gradually, and is superadded, as it were, to the lightning pains and other symptoms which characterise the first stage of the disease. In some cases, the inco-ordination has been in existence for a considerable time before the true nature of the defects which it produces is appreciated by the patient.

When the inco-ordination is slight, it may only be noticed when the guiding influence of vision is removed. The patient finds, for example, that if he gets up in the night and attempts to walk across the room in the dark, he is apt to stagger and knock himself against surrounding articles of furniture; or that when he is washing his face in the morning, the eyes being closed to prevent the entrance of soap, he is apt to fall forwards against the washing-stand.

When the inco-ordination is very slight, the patient may have no difficulty in walking about in daylight; but even at this stage of the disease, he may feel insecure when he attempts to cross a crowded thoroughfare or to walk on a slippery surface. He says that he has lost his nerve; and no doubt the difficulty in the early stages is partly due to mental or psychical causes; in other words, to a feeling of insecurity, which is really an indirect result (although the patient may not know it) of the inco-ordination. Most of us whose co-ordination is perfect have no doubt experienced this psychical form of inco-ordination in attempting to walk over a narrow plank, in descending precipitous rocks, etc. Under such circumstances, many people lose their nerve; their knees tremble and shake, and their move-

ments become uncertain and unsteady; in short, a form of acute (psychical) inco-ordination is developed. It is no wonder, then, that patients who are suffering from locomotor ataxia may find their inco-ordination greatly increased in much less dangerous positions, and that they may hesitate to place themselves in situations which at one time would have caused them no concern.

As the disease advances, the inco-ordination becomes more marked and the characteristic gait is developed.

When the ataxia involves the upper limbs, the fine (highly specialised) movements (such as those concerned in writing, picking up a pin, buttoning the trousers) are usually first disordered; as the ataxia becomes more marked the coarser movements (such as that concerned in lifting a glass of water to the mouth) become unsteady and inco-ordinate. Even when the inco-ordination is slight, the patient may be unable to direct the finger, when the eyes are closed, with steadiness and certainty to the tip of the nose or to the mouth.

Before describing the characters of the ataxia in its fully developed form, it may perhaps be advantageous to discuss the pathological physiology of the inco-ordination and to describe the methods by which we test the co-ordinating mechanism in the living man.

Pathological physiology of the inco-ordination of locomotor ataxia.

—A healthy man is able to stand with the eyes closed when the feet are placed close together, and, with a little effort, to balance himself on one leg when the eyes are closed. The muscular contractions which are necessary for the maintenance of the erect position under such circumstances are regulated:—(1) partly *under the influence of the Will*, and (2) partly *reflexly* (i.e. through the spinal cord) by means of peripheral impressions which pass to the spinal cord and brain from the skin of the soles, and from the muscles, tendons, ligaments and joints of the lower extremities and of the trunk.

I omit any reference to the semicircular canals, for this mechanism (the integrity of which is so essential for the maintenance of equilibrium and the erect position and the perfect co-ordination of movement) is not deranged in cases of locomotor ataxia.

By means of these ingoing impressions, the sensorium is kept informed:—(a) of the relationship of the feet, and therefore of the body as a whole, to the ground; (b) of the way in which the

muscles are contracting; and (c) of the direction in which the limbs are being moved, of the condition of the muscles, tendons, ligaments and joints, and of the relationship of the joint surfaces to one another. In other words, these ingoing impressions (1) supply the sensorium with the information which is required for the *voluntary* co-ordination of movement; and (2) send to the spinal cord stimuli which unconsciously (that is to say *reflexly*) throw into action and regulate the co-ordinating mechanisms which it contains.

A certain amount of co-ordination must take place in the grey matter of the spinal cord, and this co-ordinating mechanism in the spinal cord is in communication with, and capable of being put into action by:—(a) reflex impulses which enter the cord through the posterior roots; and (b) volitional impulses which pass to the grey matter of the cord by the fibres of the pyramidal tracts.

Inco-ordination may of course result from defective or badly arranged impulses reaching the spinal co-ordinating mechanism through the fibres of the pyramidal tract. But this (cerebral) form of inco-ordination does not concern us now. I see no sufficient reasons for supposing that the inco-ordination of locomotor ataxia is cerebral, i.e. due to badly arranged motor impulses passing to the spinal co-ordinating mechanism through the fibres of the pyramidal tracts—irrespective of the defective arrangement of voluntary motor impulses which must necessarily result from defective information being supplied to the cerebral centres by the lesion in the afferent (sensory) channels, i.e. in the spinal cord and peripheral nerves.

It is probable that the inco-ordination of locomotor ataxia is partly due to interference with the ingoing (sensory-producing and reflex-producing) impulses, as they pass through the postero-external columns of the cord.

But further, the lesion in the postero-external columns of the cord may perhaps disturb the spinal co-ordination of movement in another way, namely, by interrupting the connections between the grey matter (nerve cells) and nerve fibres of adjacent segments. It is probable that commissural fibres which serve to connect the grey matter of one segment of the cord with the grey matter of adjacent segments pass through the postero-external columns; and that these commissural fibres are concerned in the spinal co-ordination of movements. According to this theory, reflex impulses (and the same statement applies to voluntary motor impulses, or rather to some of the voluntary motor impulses, which enter the grey matter by the fibres of the pyramidal motor tract) must, in order to become co-ordinated in the cord, pass through the postero-external column, so that they

may form connections with nerve cells at levels above or below their point of entrance into the grey matter. According to this view, some of the reflex impulses (and some of the voluntary motor impulses which enter the grey matter of any given segment by a fibre of the crossed pyramidal tract) must, before passing out of the cord through the fibres of the anterior nerve roots, leave the grey matter and pass through the commissural fibres of the postero-external column, in order to again reach the grey matter of the cord at a higher or lower level.

This view supposes that the co-ordinating mechanism in the grey matter of the cord consists of nerve cells and fine fibres; that the nerve cells and fine fibres connected with any given movement are not necessarily, nor indeed likely, to be all situated in the same spinal segment; that the nerve cells and fine fibres of adjacent segments are probably connected together, not only by fibres and fibrils of the grey matter, but by larger and longer fibres, which pass from one segment to adjacent segments through the postero-external columns.

Further, we may suppose that this co-ordinating mechanism in the grey matter of the cord may be acted upon by impulses which pass into the grey matter either from (a) the posterior roots, or (b) the pyramidal tracts.

The importance of this connection of the fibres of the pyramidal tracts will be more apparent when I come to speak of the inco-ordination of Friedreich's ataxia.

The muscular movements concerned in the act of walking are regulated in the same manner, partly by voluntary, and partly by reflex motor adjustments, under the guidance of the ingoing impressions which pass from the lower extremities to the brain and the spinal cord. The reflex (unconscious) regulation probably plays a more important part in regulating the mechanism concerned in the act of walking than in the act of balancing the body in the erect position.

Now, we have seen that the lesion in locomotor ataxia is situated on the sensory side of the nerve apparatus. The inco-ordination of locomotor ataxia seems to be chiefly due to the fact that ingoing impressions are interfered with or interrupted, and partly perhaps to the interference with the commissural (co-ordinating) mechanism in the posterior columns to which I have just referred.

The ingoing impressions which proceed partly from the skin of the soles, ligaments, joints, bones and muscles of the lower extremities, pass to the spinal cord through the sensory branches of the cerebro-spinal nerves and the posterior roots, and upwards to the brain through the sensory tracts in the cord.

In consequence of the lesion in (a) the postero-external

columns of the cord (through which these ingoing impressions pass), and (b) the peripheral sensory nerves (more especially the fine sensory nerves in the muscles, the integrity of which is essential for the reception of muscular sense impressions), (1) the sensorium is not informed, or is incorrectly informed, of the condition of the lower extremities (of the relationship of the feet to the ground and of the way in which the muscles are contracting, etc.), and the voluntary co-ordination of movement is interfered with, though the inco-ordination can to a large extent be prevented by the information afforded by the sense of sight¹; and (2) the reflex stimulation and regulation of the spinal co-ordinating mechanisms connected with the lower extremities is interfered with.

Hence the inco-ordination.

But patients who are affected with locomotor ataxia can, unless the inco-ordination is very extreme, maintain the erect position when the eyes are open and the feet are placed wide apart, and can walk (with an inco-ordinate gait) provided that they keep their eyes on their feet and on the ground in front of them.

The information which the sensorium receives through the sense of sight compensates, in some degree at least, for the want of information received through the ingoing nerve apparatus of the lower extremities. The information which the sense of sight affords is sufficient to enable the ataxic patient to stand when his base of support is sufficiently wide; and to walk, though the movements of the lower extremities (as seen by the characteristic ataxic) gait are very imperfectly co-ordinated.

But if the ataxic patient is made to close his eyes, he is unable to maintain the erect position and he can no longer walk, for the information which he receives from the lower extremities is insufficient to enable him to perform the necessary muscular adjustments. Indeed, if the lesion is advanced and the muscular sense completely abolished, he may get no information at all from the lower extremities. Under such circumstances, he is absolutely ignorant of the condition of his muscles and of the position of his lower extremities; he is no longer able to stand, and in his attempts to walk, when supported and with his eyes closed, he may throw his legs wildly about in a most disorderly but it may be very vigorous manner.

I repeat that the inco-ordination of locomotor ataxia seems to be *partly* the result of the lesion in the posterior columns of

¹ The fact that the inco-ordination in cases of locomotor ataxia is markedly less when the eyes are open and fixed on the ground and on the feet seems to me to show conclusively that the ataxia is in part at least due to the lack of guiding impressions passing to the brain through the spinal cord, that is to say, to a lesion on the afferent side of the spinal nerve apparatus.

the cord, which deranges the spinal co-ordinating mechanism and interrupts the upward passage to the sensorium and cerebellum of ingoing impressions (which pass into the cord through the postero-external columns and upwards to the brain and cerebellum through the postero-internal columns); and *partly* the result of the lesion in the sensory nerve terminations at the periphery (muscles, tendons, ligaments, joints and skin of the lower extremities) which prevents the reception, so to speak, of the ingoing sensory and reflex impulses which are essential, when the sense of sight is removed, for the satisfactory co-ordination of muscular movements in the lower limbs.

This explanation of the ataxia is not accepted by all authorities. According to some, the ataxia is motor and not sensory, cerebral and not spinal. Loss of power in the opponent muscles, excessive contraction of the opponent muscles, an altered condition of the muscular tonicity, and a badly arranged discharge of voluntary motor impulses from the cerebral cortex have all been suggested as the fundamental causes of the ataxia. I cannot see my way to adopt any of these explanations, though I admit that defective grouping and action of the muscles and opponent muscles and defective muscular tonicity may, and undoubtedly do, all result from the lesion on the ingoing side of the nerve apparatus. The defects on the sensory side of the nerve apparatus, together with the defective arrangement of outgoing (voluntary and reflex) motor impulses in the *spinal cord* which results from interruption of the commissural connections in the posterior columns, afford, in my opinion, a satisfactory and sufficient explanation of the ataxia. I see no reason to suppose that the inco-ordination is due (as Jendrassik and Raymond have suggested) to a lesion (defective commissural connections) in the cortex of the brain.

Method of testing co-ordination in locomotor ataxia.—Our tests for inco-ordination are based upon these physiological considerations. When a patient comes before us who is supposed to be suffering from locomotor ataxia, we test the co-ordinating mechanism of the lower extremities by observing:—(1) Whether he is able to balance the body in the erect position (a) with the eyes open, and (b) with the eyes closed; (2) The way in which he walks, (a) with the eyes open, and (b) with the eyes closed; and (3) The way in which the muscular movements of the lower and upper extremities are performed when he is lying in bed, (a) with the eyes open, and (b) with the eyes closed—for it is of course necessary to test the co-ordinating mechanism of the upper as well as of the lower extremities.

Romberg's Test.—In testing the patient's power of balancing himself in the erect position, we begin with a coarse test (we

observe whether he can stand steady with the eyes open and the feet wide apart), and gradually make the test more and more difficult, narrowing the base of support by bringing the feet close together; and finally, if this test is satisfactorily performed, we see whether he can balance himself on one leg alone. We then repeat the tests with the eyes closed. If the patient can perform the final and most delicate test, i.e. if he can balance himself on one leg with the eyes closed, it is certain that there is no inco-ordination in the lower extremities.

The significance of Romberg's sign.—Inability to balance the body in the erect position when the eyes are closed is not pathognomonic of locomotor ataxia; it may result from any lesion which interferes with the upward conduction of ingoing impressions from the lower extremities; further it may be the result of simple nervousness, of muscular weakness and of inco-ordination. Nevertheless, it (Romberg's sign) is highly suggestive and characteristic of the ataxic stage of tabes dorsalis; for locomotor ataxia is the disease *par excellence* in which ingoing impressions, and especially the muscular sense impressions, are interfered with or impaired, and in which, at the same time, the muscular movements of the lower extremities are inco-ordinate.

Inability to *maintain* the balance of the body in the erect position may be due to loss of sensibility in the skin of the soles. A patient who does not feel the ground at all in consequence of complete anæsthesia of the soles is unable to stand securely (though the guiding sensations which he receives from the semicircular canals are usually sufficient to enable him to do so); he may be able to stand securely for a time, but once his balance is upset, he has difficulty in recovering himself. So long as he keeps quite steady, he may, by the aid of the ingoing impressions which he receives from the muscles, ligaments and joints of the lower extremities, be able to keep his muscles in the same position as when he started; but once his balance is upset, he is (unless the semicircular impressions are sufficient) unable to recover himself; for, however perfect his co-ordination, he has lost his bearings and he is unaware of the exact relationship of his feet to the ground. He consequently does not know (once his equilibrium is upset) the muscular adjustments which are, under the circumstances, required, though he would be perfectly able to make these adjustments if he did know them.

The inability to stand erect under such circumstances is not the result of inco-ordination. The inability to stand erect with the eyes closed is, in cases of locomotor ataxia, due to inco-ordination and loss of the ingoing impulses (other than tactile impressions) which regulate the co-ordinating mechanism. This is shown by the fact that in some cases in which the inco-

ordination is very marked, and in which the patient is quite unable to stand erect with the eyes shut, there is no discoverable or appreciable defect in the sensibility of the skin of the soles.

Testing the walking power.—We next test the patient's power of walking. We first make him walk in a straight line with his eyes open, and as he does so, we particularly note his gait. If this test fails to bring out the inco-ordination, we next direct him to walk backwards with his eyes open. This is a more severe test of his co-ordinating power. Finally, if he performs this test satisfactorily, we make him walk forwards with his eyes closed. If he performs this test satisfactorily, we conclude with certainty that there is no inco-ordination. You must remember of course that many healthy persons walk unsteadily when their eyes are closed, especially if they are uncertain of their surroundings; but they do not stagger about as patients affected with locomotor ataxia do. A healthy person can walk steadily forwards for a few paces with his eyes closed, provided that the floor is perfectly level and he knows that there is nothing in his way; in other words, provided that he feels perfectly secure of his surroundings and knows that he is not likely to knock up against anything. A patient who is ataxic cannot do this.

The gait.—The gait of patients affected with locomotor ataxia, in its fully developed form, is very striking and characteristic. In typical cases, the patient usually keeps his eyes fixed on the ground in front of him, for he requires to keep the sensorium informed, by the aid of sight, of the position of his feet and of his relationship to the ground. If you make him look up into the air, the inco-ordination becomes more marked; in fact, in advanced cases, he may then refuse (for he knows that he is unable) to walk at all.

A patient affected with locomotor ataxia usually walks in a straight line; he does not reel from side to side like a drunken man or a patient suffering from cerebral disease. As Duchenne long ago pointed out, the unsteadiness in locomotor ataxia is the result of the condition of the legs (or to speak accurately, of the spinal cord) rather than of the head. When the ataxia is marked, the patient usually has recourse to the help of a stick or sticks. The steps are taken slowly and deliberately, but the movements of the legs are irregular and jerky; the knees are kept stiff; the feet are often raised too high, and jerked forward in a sudden and uncertain way; and the heels are brought

suddenly to the ground with a stamp. Hence the term 'stamper's gait,' as the Germans call it. There are of course differences in individual cases, but that is the characteristic gait. You must all have noticed persons walking about the streets of Edinburgh who present this gait. For the past fifteen years I have observed one gentleman in particular—I have never seen him professionally—who presents the characteristic gait of locomotor ataxia in a marked degree. He does not seem to be getting any worse; but his facial expression is anxious and worn. I suspect, if I had the opportunity of questioning him, I would find that he suffers from the lightning pains which are so characteristic of the disease. I have noticed a worn and anxious expression in many cases of locomotor ataxia; but it is by no means always present. In some of the cases which have come under my notice in which the lightning pains were most severe, they were not permanently stamped in this way, as it were, upon the countenance.

When a patient affected with locomotor ataxia is told to walk, he often seems to take some time to steady himself before he starts; and if when walking he is suddenly told to halt, he seems to have some difficulty in stopping himself, i.e. he takes some time to pull himself up.

Impairment or loss of the muscular sense.—Impairment or loss of the muscular sense is an important feature of many cases of locomotor ataxia, and is in all probability the chief cause of the inco-ordination. In those cases in which the muscular sense is completely lost, the patient is absolutely ignorant, when his eyes are closed, of the position of the lower extremities, of the way in which muscles are acting, and of the amount of voluntary motor force, so to speak, which should be discharged in order to produce a given muscular movement.

Mode of testing the muscular sense.—We test the condition of the muscular sense by observing:—

(1) The patient's power of appreciating the difference between different weights, that is to say, whether he can appreciate the different degrees of muscular force or effort which are required to raise and balance different weights.

In testing the muscles of the lower extremities, different weights are tied on to the foot and the patient is told to raise the foot from the bed and to discriminate between them. When

the muscular sense is completely lost, the patient may be unable to tell the difference between a pound and an ounce. A ready way of testing his power of appreciating resistance or weight, is to take hold of the ankle (raised half a foot or so from the bed) and to tell the patient, while you make different degrees of resistance, to raise the foot up into the air. When the muscular sense is completely lost, he may be unable to tell whether you are resisting the muscular movement gently or with all your force. He may make a powerful effort to raise the leg when a slight effort only is required, and *vice versâ*. After having made the effort, he may be ignorant of the force which he has expended—a very important argument against the view that the motor centres in the cortex of the cerebrum have anything to do with the appreciation of muscular effort.

(2) The manner in which the patient is able to perform muscular movements when the eyes are closed and the control of sight removed.

In testing the condition of the muscular sense in the lower extremities, the patient should be placed flat on his back in bed with the eyes bandaged. He should then be told to raise one leg and describe a given movement (say, a circle or a cross) in the air with his foot, or to raise the foot to a given height and then to bring the heel down slowly and accurately on the big toe of the opposite foot which is lying at rest, or upon the opposite knee. The patient who has lost the muscular sense, and who is consequently inco-ordinate in his lower extremities, is unable to execute these movements with steadiness, delicacy and precision. He raises the foot too high or too low; the movement is jerky or unsteady; he fails to make the movement with precision; the leg is brought down wide of the mark, the heel failing to come down steadily on the big toe or opposite knee as required. If the bandage is now removed from the patient's eyes and he is allowed to control the movement by the sense of sight, it will be found (if the irregularity is due to inco-ordination and not merely to muscular weakness) that the movement is much more steadily and co-ordinately performed; indeed, when the inco-ordination is not extreme, the movement may now be perfectly performed.

(3) His power of appreciating the position of his limbs (after they have been placed in different positions by the physician), his eyes being closed.

The patient's eyes being bandaged, the physician moves the leg about in the air first in this direction, then in that, and finally brings it to rest in the air (if it is placed on the bed the patient may be able to say where it is, by the sense of touch); the patient is then told to take hold of his big toe with, say, his

right hand, or to point to the place where his big toe is. When the muscular sense is much unpaired or completely abolished, he may be absolutely ignorant of the position of his foot, and indeed of the whole lower extremity. He may grope about for the foot, and may at last only be able to find it when by chance his hand comes in contact with some part of the leg, and when he is able to guide himself (the hand) by the sense of touch along to the foot. (See fig. 99.)

(4) His power of touching a given spot on the surface of his body when the eyes are closed.

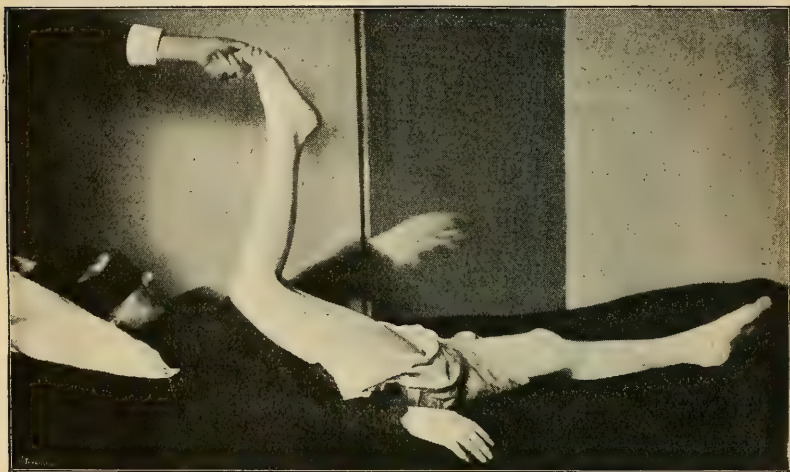


FIG. 99.—*Extreme inco-ordination and loss of the muscular sense.*

This photograph was taken while the patient was trying to find her right foot with her left hand.

She had no knowledge, when her eyes were closed and the foot raised from the bed, of the position of her foot; but she was perfectly aware that the assistant who was holding up the foot was grasping the big toe.

This test shows (a) whether the patient knows the locality of a given spot on the surface of the body; and (b) whether when his eyes are closed he is able to make the necessary co-ordinate movement to touch it.

He may be told (the eyes of course being closed) to touch the tip of his nose with his forefinger; or (and this is a much more difficult and delicate test of the condition of the muscular sense and of co-ordination) to bring the two forefingers slowly together so that their tips meet in the middle line.

(5) His power of appreciating the direction in which his limbs are moved by the passive movements effected by the physician.

This is hardly perhaps a test of the muscular sense (for the movements of the joints, ligaments and skin overlying the joints are all concerned); but it shows whether the patient receives correct impressions from the ligaments, bones, joints (and in some degree no doubt from the muscles) of the lower extremities.

The pathological physiology of the loss of the muscular sense.—As I have already told you, fine sensory nerves are extensively distributed throughout the muscles of the body. Their function is to keep the sensorium informed of the manner in which the muscles are contracting. In other words, their function is to receive and conduct to the spinal cord those ingoing impressions which are concerned in the production of the muscular sense.¹ They are intimately concerned in the co-ordination of muscular movements. The impressions which they convey to the spinal cord seem to pass up to the brain through the postero-internal columns of the cord (the columns of Goll); and in order to reach the columns of Goll they have to pass through the postero-external columns (the columns of Burdack).

But these parts—(a) the fibres of the columns of Burdack in the lower part of the cord, (b) their continuations in the columns of Goll in the upper part of the cord, and, as recent observations would seem to show, (c) the fine sensory nerves in the muscles of the lower extremities—are the very parts which are chiefly affected by the lesion in cases of locomotor ataxia.

It is the affection of these parts of the sensory (ingoing) apparatus which is the chief cause of the inco-ordination and perhaps the sole cause of the impairment or loss of the muscular sense, which are such conspicuous and characteristic features of the disease in its second or fully developed stage.

That the ingoing impressions which proceed from the muscles are much more important for the purposes of co-ordination and for the muscular sense than those which proceed from the skin is proved by the facts:—

(1) That in the first (pre-ataxic) stage of the disease, when the lesion in the posterior columns is already present and when lightning pains (and it may be anæsthesia or hyperæsthesia in the skin) are well marked, there is no loss of the muscular sense and no inco-ordination; and

(2) That in the advanced stages of some cases in which the inco-ordination is extreme and the muscular sense entirely lost, the tactile sensibility of the skin may be little if at all interfered

¹ It is doubtful if the impressions which pass inwards from the skin, ligaments, bones and joints are of much importance so far as the muscular sense is concerned; though they no doubt play an important part in informing the sensorium of the way in which passive movements of the limbs are being made.

with. Cases of this sort are rare, but they do occasionally occur; two well-marked cases have come under my own notice.

A man who has totally lost his muscular sense is absolutely helpless when his eyes are closed, so far as his lower extremities are concerned; he does not know where they are, and he does not know what he is doing with them. I have seen such a patient carried along trailing his legs behind him, uttering piteous wails, thinking every moment he was going to fall.

When the muscular sense is much impaired or completely abolished, the sensibility of the skin to touch, heat, cold and pain is usually also markedly impaired. But this, as I have already stated, is not invariably the case. In those cases in which the muscular sense is lost but in which the skin sensibility is unimpaired, it is probable I think that the lesion is situated in the sensory nerves of the muscles rather than in the spinal cord. It is very unlikely that a lesion in the cord could so seriously impair the co-ordinating power and the muscular sense without at the same time implicating the sensory fibres which proceed from the skin; while a lesion confined to the sensory nerves in the muscles might of course do so.

The condition of the muscles.—The muscles of patients suffering from locomotor ataxia are usually firm and well-nourished. Their electrical reactions are usually normal. In the early stages of the disease, a simple increase of the electrical excitability is, it is said, sometimes present. But this is exceptional. In the early stages of most cases the electrical excitability is normal or diminished (simple diminution).

In the later stages, especially when the patient is unable to walk and is confined to bed, a certain amount of muscular atrophy from disuse is almost always developed; at this stage, the electrical excitability is often in some degree diminished.

And here I would ask you to observe that locomotor ataxia, in its later stages, differs very notably as regards the condition of the muscles from spastic paraplegia. In both affections, the patient may ultimately lose the use of his limbs and be confined to bed; but in spastic paraplegia the muscles, which can no longer be acted upon by the will, are being constantly acted upon and exercised reflexly, in consequence of the exaggerated condition of the deep reflexes. It is very different in locomotor ataxia; for, as I must now tell you, abolition of the deep reflexes—the knee-jerk—is one of the most important facts in the clinical history of the disease.

LECTURE XVIII

LOCOMOTOR ATAXIA (*Continued*)

IN the last lecture, Gentlemen, we discussed the inco-ordination which is the fundamental and characteristic feature of fully developed cases of locomotor ataxia. To-day I propose to direct your attention to some of the other clinical features of the disease.

Loss of the knee-jerks.—Loss of the knee-jerks is probably the first clinical alteration, certainly it is usually the first discoverable alteration (objective sign) which the lesion of locomotor ataxia produces. In the vast majority of cases, the knee-jerks are completely abolished, when the patient first comes under the notice of the physician. In very exceptional cases, a slight knee-jerk, perhaps on one side only, can be obtained, when the other symptoms and signs of the disease are sufficiently well marked to permit of a positive diagnosis; but this is very rare.

Now, since the knee-jerks are completely abolished in the great majority of cases of locomotor ataxia, even in the earlier stages of the case, the loss of the knee-jerk constitutes *the* most important clinical sign which we possess of the disease in its early stage. I do not, of course, mean to say that you can diagnose locomotor ataxia simply by the absence of the knee-jerks. As everybody knows, loss of the knee-jerks is not pathognomonic of locomotor ataxia. The knee-jerks are lost in many other conditions. But there is no other disease of the spinal cord, excepting Friedreich's ataxia and some cases of pseudo-hypertrophic paralysis (in both of which the knee-jerks are lost), in which the muscles are firm and well nourished and the electrical reactions normal.

Mode of eliciting the knee-jerk.—It is not always easy to elicit the knee-jerk in health; for the myotatic or muscular irritability upon which the intensity of the knee-jerk depends varies considerably in different individuals. But one can almost invariably produce the knee-jerk in the healthy adult if the patient is

examined in the proper way. It is sometimes very difficult or impossible to obtain a knee-jerk in children.

When the knee-jerk is difficult to obtain, or is only very slightly marked, the patient should be seated on a high table with the bare legs dangling over the edge; he should be told to allow the legs to hang so that the muscles are completely relaxed; the ligamentum patellæ should then be sharply struck with a percussion hammer. If there is difficulty in eliciting the knee-jerk under such circumstances, the method of reinforcement must be employed. It consists in making the patient perform a powerful muscular effort. While he is seated with his legs dangling over the table, he is made to clasp his hands across the front of the chest, and is then told to pull forcibly upon the clasped hands, without, however, allowing them to be separated. Some persons while making this effort draw the knees up; the patient should be told to avoid this; while he must pull upon his hands, he must endeavour to keep the lower extremities relaxed. If the clasped hands, while they are being pulled, are kept firmly against the chest, the drawing up of the legs does not usually take place. Another method of reinforcement is to make the patient hold his breath.

The result of the reinforcement is to increase the muscular tonus all through the body. It is probable that during the performance of every powerful muscular act a wave of nerve force is sent through the whole muscular system; the excitability or tonus of muscles which are not directly concerned in the special movement which is being performed is raised. The effect is so slight that it is inappreciable externally; but it is sufficient for the purpose which we are at present considering; it so increases the excitability of the quadriceps extensor femoris that the knee-jerk (which could not previously be elicited) is readily obtained.¹

In the vast majority of cases of locomotor ataxia, it is impossible, even in the early stages of the disease, to obtain the knee-jerk by the aid of reinforcement. Hence, I repeat that loss of the knee-jerks is the most important clinical sign of locomotor ataxia, in its early stages, which we possess. I am speaking of course of true locomotor ataxia, not of the condition which is termed postero-lateral sclerosis, in which the knee-jerks are increased.

The cause of the loss of the knee-jerks.—The loss of the knee-jerk is certainly due to a lesion on the afferent side of the nerve apparatus. It is a very interesting and important fact that in

¹ Some authorities suppose that the 'reinforcement method' enables the knee-jerk to be produced by removing the cerebral control.

many cases of locomotor ataxia in which the knee-jerks are completely abolished, the superficial reflexes—the plantar reflex—can still be elicited. I have seen several cases of advanced locomotor ataxia in which the plantar reflex was markedly exaggerated. This fact seems to suggest—if we grant that the abolition of the knee-jerk is due to the cord lesion—that the nerve fibres which carry reflex impressions from (a) the skin and (b) from the

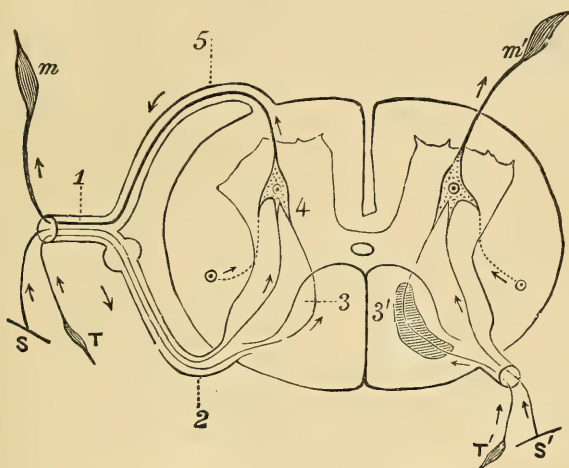


FIG. 100.

Diagrammatic representation of the reflex functions of the Spinal Segment.

The left half segment is normal. The right half represents the early stage of locomotor ataxia, the position of the lesion being shaded dark.

S, Skin from which sensory fibres pass through the common sensory motor nerve trunk, posterior root, and posterior horn of grey matter to the reflex centre (4) T, Tendon, muscle, and fascia, from which the sensory fibres pass which conduct the deep reflex movements. These fibres traverse the postero-external column.

m, Muscle supplied by the anterior root of the left half segment. The dotted line passing from the crossed pyramidal tract in the lateral column represents the inhibitory fibre.

The arrows show the course of the reflex impulse.

On the right side the deep reflex movements are seen to be arrested by a lesion in the postero-external column.

muscles and tendons pursue a different course through the cord. Several years ago I suggested the following explanation of this difference, which is diagrammatically represented in fig. 100.

Some of the fibres of the posterior root pass directly into the grey matter of the posterior horn, while others pass through the postero-external column of the cord. Now, if we suppose that the afferent fibres concerned in the production of the superficial reflexes pass directly into the grey matter, and that the afferent

fibres concerned in the production of the deep reflexes pass through the postero-external column, we have an explanation of this difference. The lesion of locomotor ataxia is, in the early stages, limited to the postero-external column of the cord; in other words, the lesion is (if this theory is correct) so situated that it will interfere with the passage of the deep, but will not interfere with the passage of the superficial, reflexes. But there is another explanation which is perhaps even more plausible. The loss of the knee-jerk is probably in some cases due to a lesion in the fine sensory nerves of the muscles and tendons. It seems certain that a lesion, either in the fine sensory nerves of the muscles and tendons, the posterior roots, or the postero-external columns of the cord, may prevent the production of the knee-jerks. You can easily understand, therefore, that if the sensory nerves in the tendon of the quadriceps extensor femoris are affected, while the plantar nerves, the posterior roots and the spinal cord are only slightly or not at all involved, the knee-jerk will be abolished, while the plantar reflex will remain.

In the lecture yesterday I told you that further information is required as to the condition of the fine sensory nerves of the muscles in cases of locomotor ataxia. Until we obtain this information we must be content to remain in doubt as to the explanation of this and some other points in the pathological physiology of the disease.

The condition of the superficial reflexes.—In some cases of locomotor ataxia the plantar reflex is abolished, in others it is preserved, indeed it may be exaggerated. I have met with several cases in which the plantar reflex was greatly exaggerated even in the advanced ataxic stage of the disease. In one remarkable case, tickling the sole was followed after a long interval by a most extraordinary contraction of the muscles in the upper part of the body; the head and upper part of the trunk were jerked up into the air by the severity of the reflex spasm, and the patient struck his head forcibly against the top of the iron bedstead on which he was lying. The jerk was so sudden and unexpected, and so violent and extraordinary that it gave me quite a fright. At first I thought that this remarkable result might be a mere accident. But it was not so. I repeated the experiment several times and on several different days, and always with the same result. The patient strongly objected to the experiment; it disturbed him greatly and produced great discomfort and exhaustion. It is difficult to explain this extraordinary alteration.

Perhaps it was due to the fact that the reflex impulse, which had great difficulty in making its way into and up through the cord, instead of passing smoothly along normal, large nerve fibres, had to pass upwards through the central grey matter, in which, as it were, it gathered head. It is possible to conceive that once it got through the diseased area of the cord, it in this way produced an exaggerated and altogether extraordinary reflex effect in the upper motor mechanisms.

I must next direct your attention to the condition of the organic reflex. In most cases of locomotor ataxia the functions of the bladder and rectum are interfered with in some degree; indeed the functions of the bladder and rectum are not unfrequently disturbed quite in the early stages of the disease.

Derangements of the bladder-reflex.—In many cases, the derangement merely consists in a deficiency of expulsive power; the patient finds that the urine is expelled slowly, less forcibly and only with a greater effort than in health. In some cases, there is ‘precipitant urination.’ In some cases, there is retention; in others, incontinence. In the earlier stages, the patient is sometimes troubled with a frequent desire to make water; this is no doubt in some cases due to reflex irritation; the ingoing (sensory) fibres of the reflex arc are irritated by the lesion; this irritation of the bladder centre may occur in paroxysms, which are probably analogous to the paroxysms of lightning pains which are so common, and of sexual excitement which are rare. In other cases, the urine dribbles away. In slight cases of this kind, the dribbling may only take place at the end of the act of micturition; but in the more advanced stages of the disease, the sphincter may be paralysed and the dribbling may be constant. In the early stages of the disease, the urinary derangement may be merely temporary; but this is exceptional. As a rule, the disturbance of the bladder persists and becomes aggravated as the disease advances. In some cases, the sensibility of the bladder and urethra is impaired.

Derangement of the rectal reflex.—Obstinate constipation is a prominent symptom in many cases of locomotor ataxia. In the later stages of the disease, the rectal sphincter sometimes becomes paralysed; but this is exceptional.

From these statements you will see that the derangements of the bladder and rectum which occur in locomotor ataxia are

somewhat different from those which characterise spastic paraplegia. These differences are easily explained by the fact that in spastic paraplegia the reflex centres for the bladder and rectum are in a state of hyperexcitability and are more easily excited than in health by reflex impressions passing from the periphery; whereas, in locomotor ataxia, peripheral impressions fail to reach or have difficulty in reaching and stimulating the reflex centres for the bladder and rectum.

Derangements of the sexual reflex.—Diminution or loss of sexual desire and power is a prominent symptom in many cases of locomotor ataxia; it may be an early symptom; it is generally present when the disease is at all advanced. In other cases, more especially in the early stages, there is increased sexual desire and occasionally (though this is rare, for the increased sexual desire is often associated with diminished sexual power, in other words, with a condition of irritable weakness of the sexual centre) increased sexual power.

In one case which came under my observation recently, the attacks of satyriasis, which consisted not merely of increased sexual desire but also of increased sexual power (for the sexual act was perfectly performed many times—four or five times—during the twenty-four hours for several days and nights in succession) occurred in paroxysms: each paroxysm, which lasted for several days, was followed by a period of complete sexual apathy and loss of sexual power, which in its turn was again succeeded by another paroxysm of furious sexual desire and enormously exaggerated sexual power.

Lightning and other pains.—Sharp, shooting pains, occurring in paroxysms, are highly characteristic of the disease. They occur in the great majority of cases; in quite exceptional cases, they are very slight or altogether absent. The lightning pains are usually developed before the ataxia, and they generally persist throughout the whole course of the disease. They are in many cases very severe and distressing; they are consequently, both from the standpoint of the patient and the physician, for they urgently demand relief by treatment, one of the most important symptoms, perhaps *the* most important, of the disease.

I have said that the lightning pains usually occur in paroxysms. Each paroxysm is made up of innumerable individual

pains. The duration of any individual pain is merely momentary; it is a flash, a stab, and it is gone; hence the term '*lightning*' pains. The duration of an individual paroxysm varies; it may last for a few hours or even for a day or more. The pains are lancinating, stabbing, darting or burning in character. In many cases, they are said to resemble the pain which is produced by the stab of a knife or the pain which would be produced by thrusting a red-hot wire into a joint. If you closely observe a patient during one of these paroxysms, you will often see that he makes a grimace or gives a jump or start when the painful stab occurs.

The skin over a part of the body in which the lightning pains are experienced is often hyperæsthetic and tender to the touch.

The paroxysms usually occur at irregular intervals; there may be days, weeks, or even months between them. In some cases, the pains are worst at night; in others, they seem to be associated with particular states of the weather, and this is no doubt one reason why they are often thought to be rheumatic. In some cases, the pains seem to be produced by excessive muscular effort or fatigue; in others by mental excitement, worry, etc.

Between the paroxysms, the patient is usually quite free of pain. In those cases in which the pains are well marked and severe, the a-neuralgic interval does not as a rule last for more than two or three weeks; in many cases, indeed, it is much shorter than this.

The pains are often, in fact usually, only felt in the lower extremities and the lower part of the trunk, i.e. the parts connected with the affected (lumbar and lower dorsal) regions of the cord. The stabbing pains which are in some cases experienced in the region of the neck of the bladder and of the rectum are especially distressing and difficult to bear. In some cases, the pains shoot round the lower part of the abdomen and thorax. The girdle sensation, which in my experience is a much less constant feature of locomotor ataxia than some writers seem to suppose, is in many cases associated with a hyperæsthetic condition of the skin; in some cases, with pains which shoot round a localised area of the trunk.

In some cases, the lightning pains are felt in the upper as well as in the lower limbs, often in the little and ring fingers; in these cases the movements of the arms may be ataxic, a condi-

tion which results from involvement of the postero-external columns in the cervical region of the cord. In exceptional cases, the pains are first or most felt in the arms. In these cases, which are very rare, the cervical region is more affected than the lumbar region. Occasionally the pains occur in the head (i.e. in the area of distribution of the fifth nerve) and neck; we have seen that the ascending and descending roots of the fifth nerve are sometimes diseased, and the peripheral fibres of the nerve itself are no doubt in some cases involved. It is only, however, in exceptional cases that the pains are referred, in any marked degree, to the head, neck and the upper extremities.

In many cases, the lightning pains are referred to the deeper rather than to the superficial structures—to the muscles and joints rather than to the skin. Painful sensations in the skin are, however, of common occurrence; they are often described as burning in character; they are usually more fixed and continuous than the true lightning pains, with which, however, they are frequently associated. The affected area of skin is often exquisitely tender to the touch. Vasomotor or trophic alterations (redness, falling out of the hair, an eruption of herpetic vesicles) are in some cases developed in those parts of the skin in which the lightning pains or burning sensations have been experienced.

In some cases, very acute pains are experienced in the position of the internal viscera (stomach, intestine, rectum, bladder, uterus, etc.). I have already mentioned the bladder and rectal pains. I shall refer in more detail to these visceral pains presently, when I come to speak of the so-called gastric and other crises.

The cause of the lightning pains.—The lightning pains are usually supposed to be due to the irritation of the posterior root-fibres as they pass through the affected (postero-external column) portion of the cord; the pain is referred, in accordance with the law of 'eccentric projection,' not to the position of the lesion, but to that portion of the periphery to which the irritated fibres are distributed. In some cases, the pains are doubtless due to an inflammatory process in the peripheral nerves themselves.

Derangements of the sensibility of the skin.—Let us now look at some of the other derangements of sensation which may occur in this remarkable disease.

Anæsthesia and hyperæsthesia.—In addition to the lightning and burning pains in the skin and the localised areas of hyperæsthesia which I have already described, the sensibility of the skin may be disturbed in a variety of ways.

Derangements of subjective sensations, such as pins and needles, numbness and formication in the lower extremities, particularly in the feet and legs, are of frequent occurrence. In many cases in which the sensibility of the skin of the soles is impaired, the patient says that he feels as if he were standing on a bag of feathers, on a soft cushion, etc.

Objective disturbances of the skin sensibility can in many cases be demonstrated. The sensibility to (a) touch, (b) pain, and (c) heat and cold, may be impaired or lost, collectively or individually. Localised areas of anæsthesia can sometimes be demonstrated in parts of the skin which were, a short time previously, the seat of lightning pains, burning sensations, etc. The anæsthesia is usually localised; it is very rarely diffused all over the lower extremities; it is generally more marked in the feet and in the legs than in the thighs, and particularly in the soles. In some cases, areas of hyperæsthesia or anæsthesia may be detected over the lower part of the abdomen, and especially over the scrotum, penis and perineum.

It is important to remember that, although the disturbances of sensation which I have just described are usually present in the ataxic stage of the disease, they are not invariable. In several cases in which lightning pains, ataxia and loss of the muscular sense were very marked, I have been absolutely unable to discover any objective signs of sensory disturbance in the skin (any anæsthesia or hyperæsthesia). In very rare cases, in which the ataxia is extreme, the muscular sense entirely lost, the knee-jerks absent and the Argyll Robertson condition of the pupil well marked, the lightning pains and other disturbances of sensation are entirely, or almost entirely, absent.

In the upper extremities numbness is not unfrequently complained of in the hands and fingers, more especially in the little and ring fingers (area of distribution of the ulnar nerve), but definite objective anæsthesia is rare.

Delayed sensations.—In some cases, a pinch or prick or touch is not felt by the sensorium with the rapidity with which it is

felt in health. In the remarkable case to which I referred a few moments ago (see page 310), tickling the sole was followed after a long interval (many seconds) by a very remarkable contraction of the muscles of the upper part of the body. In that case, the sensation, like the reflex muscular contraction, was enormously delayed. The ingoing impression seemed to have the greatest difficulty in making its way through the diseased and degenerated cord.

Erroneously projected sensations.—In some cases, a touch or pinch is felt but erroneously projected by the sensorium; it reaches the brain and a conscious impression is produced, but instead of being referred to the part which is touched, it is referred to some adjacent part. For example, you may touch the toe and the patient may tell you you are touching the leg or thigh. In very rare cases the erroneous projection is still more marked; the touch may actually be referred to some part of the opposite leg. In rare cases a double sensation (of touch and pain) results from a single impression.

Derangements of the muscular sense.—In describing the inco-ordination I have already referred in detail to the diminution or loss of the muscular sense, which in many cases is an important striking and clinical feature of the disease. I happen to have in Hospital at the present time a very interesting case in which the muscular sense is completely abolished, but in which the sensory functions of the skin are scarcely if at all impaired. The details of the case are briefly as follows:—

Case of extreme ataxia and loss of the muscular sense; no impairment of the sensibility of the skin.—The patient is an unmarried woman, aged 36. The ataxia is extreme; the knee-jerks are lost; the Argyll Robertson condition of the pupil is well marked; the muscular sense is entirely abolished; but there has scarcely been any pain and there is no objective defect (anæsthesia or hyperæsthesia) in the skin sensibility. While lying in bed, the patient is able to move the limbs with a fair amount of force and precision, so long as she sees them; the legs are thin and wasted, but she is naturally spare. She is absolutely unable to stand even with the eyes open and the feet wide apart; and even when supported (held up) on each side, she walks with the greatest difficulty. When she is made to walk with her eyes closed, a nurse holding her up on each side, she throws her legs about in the most irregular and violent manner; she does not know where

the legs are, how she is moving them, or how she ought to move them; the legs and feet seem to get twisted and mixed up. If, when the patient is lying on her back in bed with her eyes bandaged, the leg is raised and placed in some position in the air, the patient is quite ignorant of the position of the foot. [The photograph which is reproduced in fig. 101 shows this in a very striking way. It was almost instantaneous, and was taken while the patient was groping about for her foot. The hand which was in movement is blurred.] The patient could

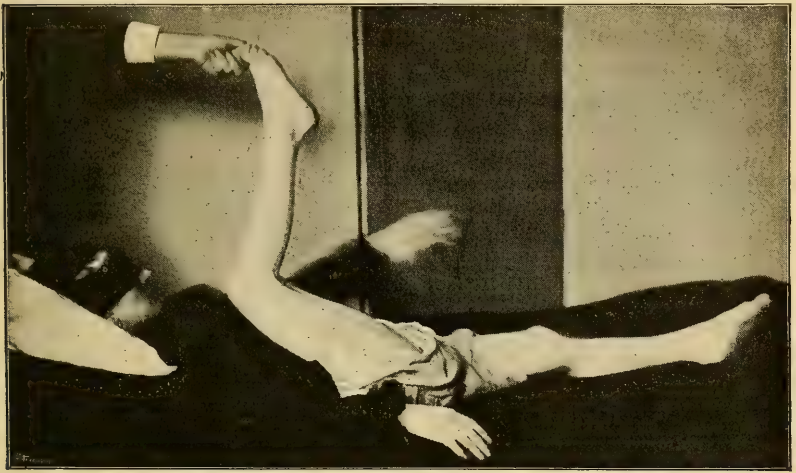


FIG. 101.—*Extreme inco-ordination and loss of the muscular sense.*

This photograph was taken while the patient was trying to find her right foot with her left hand.

She had no knowledge, when her eyes were closed and the foot raised from the bed, of the position of her foot; but she was perfectly aware that the assistant who was holding up the foot was grasping the big toe.

only find the foot if she happened to bring the hand, accidentally as it were, in contact with some part of the limb; she was then able to guide it along the leg to the foot by the sense of touch. I will return to this case when I come to speak of the diagnosis. The diagnostic problem which had to be solved was, Were the symptoms due to organic disease or to functional disturbance? The point which I wish now to emphasise is this, that the muscular sense was entirely abolished, while there was little or no obvious defect in the sensibility of the skin.

The common sensibility of the muscles, bones and joints.—The sensibility of the muscles to pain is sometimes also impaired or abolished. The patient may feel no pain when the muscles are forcibly pinched or when they are stimulated by a strong faradic current.

In some cases, the anæsthesia involves all the deep-seated tissues of the limb (the joints, bones, periosteum, etc.). It is remarkable that in many of the cases in which Charcot's joint affection is developed, there is absolutely no pain although the joint surfaces are undergoing rapid disorganisation.

I must next describe the ocular symptoms; they are numerous and important.

Abolition of the pupil reflex to light.—This symptom or rather sign was first described by Dr. Argyll Robertson. It has consequently been termed *the Argyll Robertson condition of the pupil*. It consists in the fact that the pupil fails to contract to the stimulus of light, but contracts as in health during an act of accommodation, i.e. when the patient rotates the eyeballs inwards and 'fixes' a near object. It is a very important sign from a diagnostic point of view. Though less constant than the loss of the knee-jerks, it is present in the great majority of cases of locomotor ataxia, and it is usually developed in the early (pre-ataxic) stage of the disease. Further, it is an objective sign which can be seen by the physician and which cannot be manufactured by the patient. And lastly—and this is a point of great importance—it is, so far as I know, never developed in purely functional conditions. It is a sign of organic disease, and in particular of two organic diseases (which are closely allied pathologically and etiologically)—for it rarely occurs in any other conditions—viz., locomotor ataxia and general paralysis of the insane.

The cause of the Argyll Robertson condition of the pupil.—The Argyll Robertson condition of the pupil is probably due to a lesion on the afferent (sensory) side of the reflex arc. The lesion is probably situated between the point at which the nerve fibres which conduct visual impressions pass off (in their upward passage to the visual centre), and the centre for the pupil reflex in the upper part of the pons Varolii. This seems to be proved by the fact that a beam of light which is thrown into the eye produces a visual sensation, but fails to produce contraction of the pupil; in other words, the stimulus passes to the visual

centre, but does not reach the pupil reflex centre. I am taking it for granted that the lesion is situated on the sensory side of the reflex arc. Of course the block may be situated in the pupil reflex centre itself; but this is, I think, less likely; for the accommodation centre which lies in close juxtaposition to the pupil reflex centre is unaffected. Further, analogy (the fact that the other symptoms of locomotor ataxia are on the sensory side of the nerve apparatus) is opposed to the view that the reflex centre itself is the seat of the lesion.

But these theoretical considerations are of no great moment. The practical points—and they are of the greatest importance—are these:—that the pupil reflex to light is lost in the great majority of cases of locomotor ataxia; that the Argyll Robertson sign is usually developed at an early stage of the disease; and that as a diagnostic sign of locomotor ataxia it is only secondary in importance to the loss of the knee-jerk.

Failure to dilate when the stimulus of light is removed.—In some cases of locomotor ataxia, the pupil not only fails to contract when a ray of light is thrown into the eye, but fails to dilate when the stimulus of light is removed.

Failure of the pupil to dilate under sharp stimulation of the skin.—In some cases of locomotor ataxia, the pupil fails to dilate when a sharp stimulus is applied to the skin. In health, powerful stimulation of the skin (a sharp prick with a pin, a pinch, or the sudden application of a powerful faradic current) is followed by reflex dilatation of the pupil. Fright produces the same thing. You have all, no doubt, seen a frightened cat, and you may perhaps have noticed how widely dilated its pupil becomes. Well, we take advantage of this fact in disease. If the sensory stimulus is unable to pass through the peripheral nerves or spinal cord, this reflex dilatation of the pupil does not occur. Absence of reflex dilatation of the pupil is not a sign of much importance. It is much less important from a diagnostic point of view than the loss of the light reflex. It merely shows that the upward conduction of sensory impressions from the portion of skin which is stimulated is interrupted (either in the peripheral nerves or in the spinal cord). It occurs in many other conditions besides locomotor ataxia.

Alterations in the shape and size of the pupils.—In some cases of locomotor ataxia, the pupils are unequal in size; in others, they are irregular in shape. These alterations are not common and are of no great significance. They are much less common in

locomotor ataxia than in general paralysis of the insane. In that disease inequality of the pupils and irregularity of the pupil are, as I shall afterwards have to tell you, of some diagnostic importance.

In some cases of locomotor ataxia, the pupils are extremely contracted. This pin-point condition, or *spinal myosis* as it is technically termed, is probably due to involvement of the sympathetic fibres in the cilio-spinal (upper dorsal and lower cervical) parts of the cord.

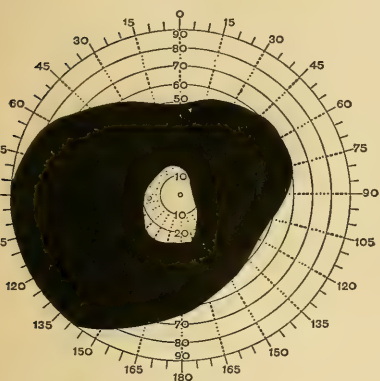
In other cases, but this is much less common, and is chiefly seen in those cases in which there is optic atrophy, the pupils are abnormally dilated (*mydriasis*).

Paralysis of the ocular muscles.—In a few cases of locomotor ataxia the accommodation becomes paralysed, but in my experience this is exceedingly rare. A much more common condition is temporary paralysis of some one or other of the external muscles of the eyeball. When this *disassociated paralysis*, as it has been termed, is slight, the effect is merely double vision (diplopia) without any appreciable or very distinct alteration in the position of the eyeball. When the paralysis is more marked there may be strabismus. In those cases in which the levator palpebræ superioris is paralysed there is ptosis. These ocular paralyses, which usually occur in the early (pre-ataxic) stage of the disease, are in the great majority of cases temporary. In rare cases, the ocular paralysis is more lasting or permanent; in some cases of this kind, the paralysis is due to a coarse syphilitic lesion, a gumma or localised meningitis at the base of the brain, which involves the third or the sixth nerves; in others, to a neuritis involving the trunk of the affected nerve; in others, perhaps, to a lesion of the nerve nucleus. But, as I shall have to tell you when I come to speak of the complications, gummata and other syphilitic lesions (if we except general paralysis of the insane) are rarely associated with locomotor ataxia.

In some cases, all the external or internal muscles of the eyeball are paralysed. These conditions, ophthalmoplegia externa and interna, as they are technically termed, are usually due to a nuclear lesion. They are highly suggestive of a syphilitic lesion. They are, I think, probably the result of an associated syphilitic lesion rather than the (direct) result of the lesion which is the pathological substratum of locomotor ataxia.

Optic atrophy and amblyopia.—The alterations in the ocular apparatus which I have up to this point described are not of any great importance, so far as the prognosis is concerned; though some of them (the Argyll Robertson condition of the pupil and the temporary paralysis of the ocular muscles—especially the former) are of great importance from a diagnostic point of view. But there is one alteration, viz., optic atrophy, which, from the patient's point of view, is of the greatest importance. In a certain proportion of cases of locomotor ataxia,

LEFT.



RIGHT.

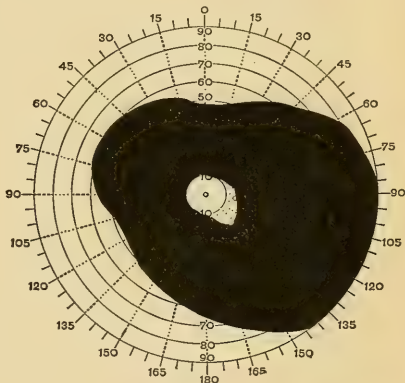


FIG. 102.—*Constriction of the fields of vision in a case of pre-ataxic tabes.*

The optic discs were markedly atrophied, the Argyll Robertson pupil was well marked and the knee-jerks were abolished. There were no other symptoms, in particular no lightning pains.

the optic nerves are affected with grey atrophy. The result is loss of vision; the acuity of vision becomes impaired, and the fields of vision, both for white and colours, but especially for green and red, become constricted. The constriction of the fields is with rare exceptions concentric (see fig. 102); and the degree of constriction is usually fairly equal in the two eyes, but this is by no means always the case. In rare cases, while the loss of vision is complete or almost complete in one eye, central vision (the acuity of vision) is almost normal in the other. The same alteration is occasionally observed in general paralysis of the insane. A remarkable illustration (with post mortem) came

under my notice a short time ago. (See fig. 103.) In some cases, the power of seeing red and green is completely lost (dyschromatopsia). In the great majority of cases, the optic atrophy and resulting amblyopia, once they are developed, slowly and gradually advance; complete blindness may ultimately be produced. Fortunately for the patient, optic atrophy is, comparatively speaking, a rare result of the disease.

The optic atrophy is usually developed in the early (pre-ataxic) stage of the disease; and it is a remarkable fact, which is

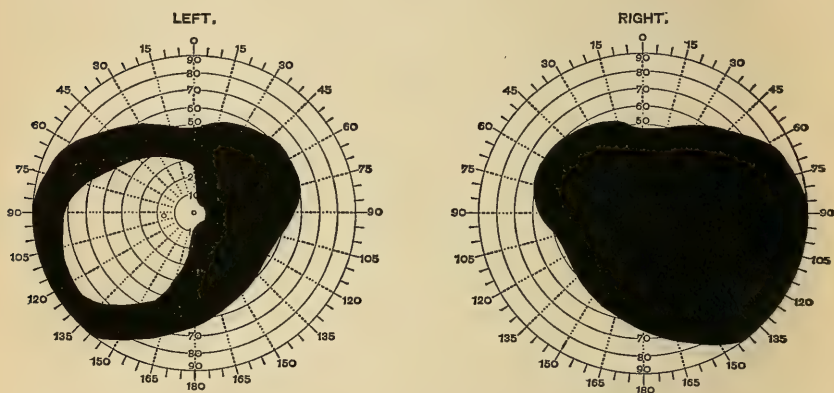


FIG. 103.—*Constriction of the fields of vision in a case of optic atrophy and general paralysis of the insane.*

Vision was *nil* in the right eye; the acuity of vision in the left eye was normal; the half of the field in the left eye was markedly constricted. The visual defect was suggestive of a lesion of the optic chiasma; but the examination of the optic nerves after death showed a primary grey atrophy, complete in the right eye and incomplete in the left.

of considerable importance for the purpose of prognosis, that in many of the cases in which the optic atrophy is developed early (together with lightning pains, loss of the knee-jerk, and perhaps the Argyll Robertson condition of the pupil) the ataxic symptoms are never developed, or are more slowly developed than in the average run of cases. It has indeed been suggested that the optic atrophy exerts a retarding influence upon the development of the ataxic symptoms.

In some of these cases, gastric crises are associated with the optic atrophy and the other symptoms and signs characteristic of the pre-ataxic stage of the disease. In other cases, the

peculiar lesion of the bones and joints, which we term Charcot's joint disease, is also present.

Charcot was of opinion that optic atrophy, the joint affection and the gastric crises are more apt to be developed in the early stages of female than in the early stages of male cases. This is not my experience. I have only once seen optic atrophy in a woman affected with locomotor ataxia. In explanation of this statement I should add that I have seen very few female cases of locomotor ataxia. It would appear that female cases are more common in France than in this country.

The atrophy is a grey atrophy; the edges of the disc, which retains its oval shape and its normal size, are sharply defined; the disc loses its transparency, but has not the chalky whiteness and the want of definition at its margins which are characteristic of post-neuritic atrophy. Further, the retinal arteries and veins are not markedly constricted, and the white lines, which in cases of post-neuritic atrophy can often be seen extending along the edges of the vessels, are not present. In short, the atrophy is primary and degenerative in character. The blindness is usually progressive and incurable. But this is not always so. Several years ago, I reported a case of pre-ataxic tabes with optic atrophy, in which the condition of vision improved in a most remarkable way under treatment. A few other cases of the same sort have been recorded.

Very exceptionally, flashes of light are experienced by the patient in cases in which the optic nerves are degenerating. The flashes of light may occur in paroxysms which are probably analogous to the paroxysms of lightning pains.

Auditory defects.—Impairment or loss of hearing, noises in the ears and tinnitus—symptoms indicative of affection of the auditory nerve or of the internal ear—are developed in a certain proportion of cases of locomotor ataxia. Slight defects are probably not uncommon; marked derangement of hearing is certainly very rare.

In some cases, the active auditory symptoms (the noises in the ears and the vertigo with which the tinnitus is not unfrequently associated) occur in paroxysms. In one well-marked case these paroxysms were relieved by antipyrin; in another by phenacetin—remedies which are often very effective in relieving the lightning pains.

Visceral crises.—Let me now refer to the visceral crises. I have already more than once stated that the functions of the stomach are sometimes acutely deranged in this disease. But the stomach is not the only organ which is affected. It is, however, more frequently and more severely affected than any other organ.

The term *gastric crises* has been given to these paroxysmal attacks of stomach derangement. They are characterised by severe pain in the region of the stomach, radiating to the back, chest, etc., and by urgent vomiting and retching which may be unattended by nausea or any marked derangement of the tongue. The gastric symptoms occur in paroxysms and are usually developed suddenly and without any obvious exciting cause; in many cases they subside as rapidly and unexpectedly as they appeared. The paroxysm may continue for several days. Between the paroxysms the gastric functions may be perfectly performed. The vomited matters in many cases consist of glairy mucus, in some cases mixed with bile. In rare cases, the vomited matters may contain blood, the presence of which may give rise to the suspicion of a gastric ulcer.

During the attacks, the pulse becomes greatly accelerated and a condition of profound collapse is not unfrequently developed.

These gastric crises are usually developed in the early stage of the disease, in association with the other characteristic symptoms (lightning pains, loss of the knee-jerks, the Argyll Robertson condition of the pupil) of the pre-ataxic period. In some of these cases, optic atrophy and Charcot's joint affection are also present. In rare cases of pre-ataxic tabes, the gastric crises occur without any lightning pains. Some years ago, I had several opportunities of seeing a patient who suffered at frequently recurring intervals from the most severe and pronounced gastric crises. He denied having had syphilis; his knee-jerks were completely absent; the Argyll Robertson condition of the pupil was present; and there was complete loss of sexual desire and sexual power; but there was no inco-ordination, and there had never been any lightning pains. I came to the conclusion that the case was a rare form of imperfectly developed tabes; and this opinion was afterwards confirmed by Charcot, who gave the patient a written diagnosis which I have

recently had the opportunity of seeing. I may say that the patient is in other respects in good health; able to lead an active life (shoot, fish salmon, etc.); and that his frequently recurring gastric attacks, which when I first saw him were only relieved by morphia, are now kept in check by means of phenacetin, a remedy with which at that time I was not acquainted.

The gastric crises are probably due to a lesion of the vagus nucleus or of the vagus nerve itself.

Intestinal crises, characterised by sudden and apparently causeless paroxysms of abdominal pain and profuse watery diarrhœa; and *rectal crises*, characterised by excruciating pain in the rectum, a feeling as if the rectum were distended with a red-hot ball, and by tenesmus and straining, occasionally occur.

Vesical crises, characterised by pain in the region of the neck of the bladder, and in some cases by frequent micturition and other indications of bladder irritation, are sometimes met with.

Renal crises, characterised by paroxysmal attacks of pain shooting down the course of the ureter, and closely resembling the pain of renal colic, have also been described; but no case of this kind has come under my own notice.

Hepatic crises, characterised by severe paroxysmal pain in the region of the liver which may radiate towards the right shoulder, and which may even be accompanied by slight jaundice, occasionally occur.

Laryngeal crises, apparently due to spasmodic contraction of the muscles of the larynx, and characterised by sudden and it may be great dyspnœa, cyanosis, cough, and severe pain in the neck, upper part of the spine and shoulders, and sometimes attended with vertigo, fainting or loss of consciousness, occasionally occur but are fortunately very rare. They are of great practical importance; for the patient may die during the attack. They are, so far as I know, the only cause of sudden death in locomotor ataxia (excluding, of course, angina pectoris, cardiac lesions and especially aortic regurgitation, a valvular defect which is by no means very uncommon).

Cardiac crises, characterised by paroxysmal attacks of palpitation, tachycardia, cardiac pain, shortness of breath, faintness, a feeling of impending death, etc., are occasionally met with in the course of the disease. But you must remember that palpitation is a common condition; an attack of palpitation in a person

who is suffering from locomotor ataxia may be, and in fact very generally is, due to some condition which is in no way connected with the disease. It is only when the palpitation recurs in the form of paroxysms and without any apparent exciting cause, when it is associated with the other symptoms indicative of a cardiac crisis, and when there are no indications of cardiac or aortic disease likely to produce an ordinary attack of angina pectoris, that we are justified in concluding that it is the direct result of the disease.

LECTURE XIX

LOCOMOTOR ATAXIA (*Continued*)

Vasomotor and trophic derangements.—In order to complete the clinical history of locomotor ataxia, I must briefly refer to the vasomotor and trophic disturbances. They are not common; but they are of great scientific interest, and some of them are of practical importance.

Herpetic vesicles, sometimes appearing in successive crops, are perhaps the most frequent, though by no means the most important, trophic changes which occur in the course of the disease. They are usually developed on those parts of the skin at which the lightning and other pains are prominent; the development of the vesicles often immediately follows an attack of pain. Erythematous eruptions, ecchymoses, extravasations of blood and falling out of the hair are also sometimes observed at the seat of the pains and in areas of skin which were hyperæsthetic during the paroxysms of pain. In some cases, the nails become brittle, thickened and furrowed; in others, they actually drop off, usually as the result of an extravasation of blood into the matrix. Ulcers sometimes form about the roots of the nails or between the toes. In rare cases, the teeth loosen and drop out: this 'shedding of the teeth' is not preceded by toothache; it seems to be due to trophic changes in the alveolar process of the jaw. Extravasations of blood or blisters sometimes form on those parts of the sole which are subjected to pressure and irritation, especially beneath the metatarso-phalangeal articulation of the toes. Ulcers may subsequently develop at the seat of the blisters. (See figs. 104 and 105.) In some cases, the ulcer penetrates into the deeper tissues, involves the bones and ultimately perforates through the whole thickness of the foot. In consequence of the ulceration and destruction the toe may drop off. In the later stages of the disease, bedsores occasionally develop. Trophic changes in the walls of the bladder occasionally perhaps occur, but in the great majority of cases in which the

bladder or the kidney is diseased, the inflammatory, ulcerative, or suppurative changes are due to paralysis of the bladder, septic cystitis and its results.

Lesions of the bones and joints.—In some cases of locomotor



FIG. 104.—*Perforating ulcer of the foot.*



FIG. 105.—*Perforating ulcer of the foot and loss of the great toe.*

(I am indebted to Dr. Handford for the photograph from which this figure was drawn.)

ataxia, the nutrition of the bones and joints is profoundly altered. (See fig. 106.) Professor Charcot first directed attention to these remarkable alterations in the bones and joints, and the disease of



FIG. 106.—*Patient affected with the joint lesion of locomotor ataxia.*—(After Charcot.)

the joints is now generally termed 'Charcot's joint disease.' 'Charcot' is not a term that an ignorant person readily takes up, and there is a very amusing story told of a medical student who, at his clinical examination, was told off to examine a case of locomotor ataxia with a well-marked trophic lesion of the knee-joint. He knew nothing about the disease, and as students will do under such circumstances, he tried to make up for his defec-



FIG. 107.—*Charcot's joint disease ; enormous bony thickening of each knee.*

The patient was under the care of Mr. Caird, who kindly allowed me to take the photograph reproduced in this figure.

tive knowledge by 'pumping' the patient. In answer to the question, 'What do the doctors say is the matter with you?' the patient replied, 'Oh! they say I've "got charcoal in the joint!"' Never having heard of Charcot or Charcot's joint disease, the student gravely told the examiner when he came round to ques-

tion him that the man 'had charcoal in his joint.' This little anecdote may help you to remember this very interesting condition—Charcot's joint disease.

In some cases, the joint affection is very rapidly developed. A pale, painless, non-febrile swelling, which in some cases is of great size, and which is due to œdema of the skin and subcu-



FIG. 108.—*Charcot's joint disease ; great bony thickening of the right knee.*

The patient was under the care of Mr. Miller, who kindly allowed me to take the photograph reproduced in this figure.

taneous tissues and to dropsical effusion into the joint itself, is first developed. In some cases, the swelling disappears after lasting for a certain time; in others, the affected joint becomes disorganised. It creaks or grates on movement. Bony out-growths and osseous formations in the cartilages, ligaments, and

it may even be in the adjacent muscular tissues, are formed. (See figs. 107 and 108). Finally, the joint structure becomes disorganised, the cartilages, joint surfaces and heads of the bones are rapidly absorbed (see figs. 109 and 110), producing a condition very similar to that which results from rheumatoid arthritis; but the joint affection of locomotor ataxia differs from the joint affection of chronic rheumatoid arthritis in two important particulars, viz., the acuteness with which it is developed, and the fact that it is usually quite painless. The absence of pain is a very remarkable feature of the condition. In consequence of the rapid destruction of the joint surfaces, the bones may become displaced (see fig. 111) or dislocated.



FIG. 109.

FIG. 109.—*The head of a humerus from a case of locomotor ataxia affected with Charcot's joint lesion.*



FIG. 110.

FIG. 110.—*Healthy humerus for comparison.—(After Charcot.)*

The knees, hips, shoulders and elbows are most frequently affected; the smaller joints (the wrists, ankles and the joints of the fingers and toes) are rarely involved.

The joint affection shows no tendency to repair.

Spontaneous fracture of bones.—In some cases, the bones are so brittle that they are fractured with the greatest facility. The tissues surrounding the fractured ends of the bones may become enormously swollen.

Several cases of this joint affection have come under my notice. The first occurred in a patient who was employed as a ward attendant in the Tynemouth Union Workhouse Hospital; in that case the left shoulder and the left elbow were affected;

the patient was not ataxic, but every three or four weeks he suffered from excruciating neuralgic (lightning) pains in different parts of the body. At that time, now five-and-twenty years ago, I knew nothing about the joint affection of locomotor ataxia, and

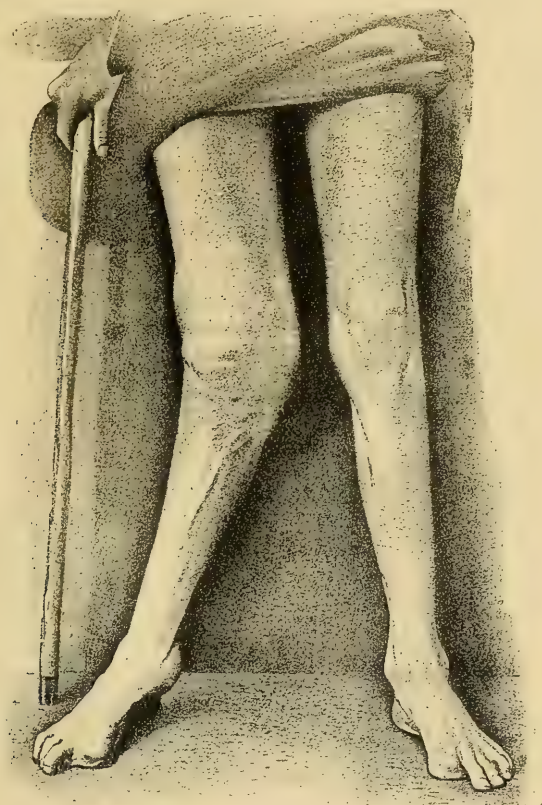


FIG. 111.—Charcot's joint disease; the right knee-joint is partially dislocated; the big toe on each foot has ulcerated off.

(I am indebted to Dr. Handford for the photograph from which this drawing was made.)

I did not recognise the true nature of the case. I saw that it was peculiar, for the joint disease, which had completely disorganised the joint surfaces and produced dislocation both at the shoulder and elbow, was absolutely painless.

The next case came under my observation in the Newcastle Infirmary. The patient, a middle-aged man, was admitted into

one of the surgical wards on account of an enormous swelling of the left thigh, which was thought at first to be a sarcoma. The thigh bone was fractured; the ends of the fractured bones could be freely moved in the middle of the swelling and made to grate and crepitate without causing the patient any pain or discomfort. The history which the patient gave us was very instructive. He was a gardener, and one day during his work his thigh bone broke with a crack. The fracture was unattended with pain. For several days after the accident he hobbled about on the broken leg, doing his work. The thigh became enormously swollen, and it was because of the swelling that he applied for admission to the Infirmary. In this, as in the last case, the patient suffered from frequently recurring paroxysms of lightning pains, and his knee-jerks were abolished; but there was no ataxia. In short, the case was a typical one of pre-ataxic tabes.

Since coming to Edinburgh, I have seen several other cases in which the joint lesion was present. In two cases, the patients were women; and I may say in passing that the joint affection seems to be, comparatively speaking, more common in women than in men. In a considerable proportion of the cases in which the joint lesion occurs, the patients also suffer from gastric crises—a fact which Dr. Buzzard has advanced in favour of his theory that the trophic centre for the bones and joints is situated in the medulla oblongata, in the neighbourhood of the vagus centre; for it is usually supposed that the gastric crises are due to a lesion which involves the vagus nucleus or the vagi nerves.

In those cases of locomotor ataxia in which the joint affection is developed, lightning pains are usually very conspicuous. Optic atrophy is sometimes associated with the joint affection. In fact the joint lesion is almost always developed in the pre-ataxic stage of the disease. Further it is important to note that in a considerable proportion of the cases in which the joint lesion, gastric and other crises, or optic atrophy are developed, the disease does not advance to the ataxic stage. Cases of pre-ataxic tabes are of great interest, and one of the reasons why they are so interesting is this, that some of the rare symptoms (joint lesions, gastric and other crises, optic atrophy, etc.) which are comparatively speaking rare in the typical and fully developed (ataxic) cases are in them (the cases of pre-ataxic tabes) comparatively speaking much more common.

In the cases which are represented in figs. 107 and 108 there was an enormous production of new bone around the affected (knee) joints; the patients were admitted to the Edinburgh Royal Infirmary under the care of Mr. Caird and Mr. Miller respectively, on account of the swelling of the knee.

Occasionally (but this condition seems very rare in this country and personally I do not remember to have seen a single example of it), the foot becomes deformed as the result

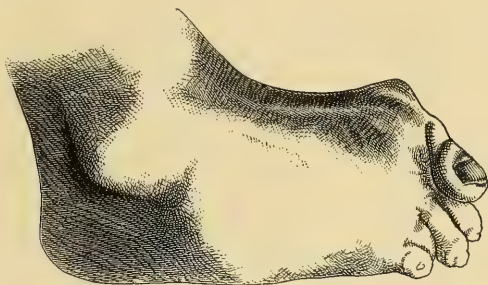


FIG. 112.—*The tabetic foot.*—(After Charcot.)

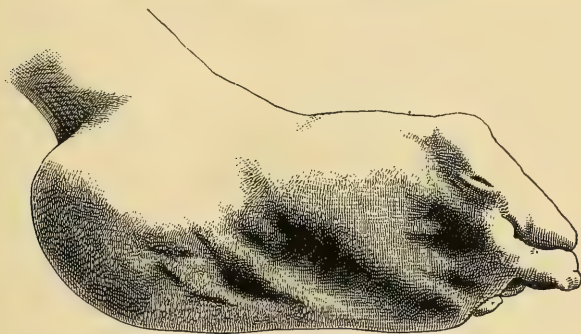


FIG. 113.—*The tabetic foot.*—(After Charcot.)

of a trophic lesion in the tarsal and metatarsal bones and joints (see figs. 112 and 113). This '*tabetic foot*' seems sometimes to be associated with the perforating ulcer to which I have already referred.

The cause of the vasomotor and trophic derangements.—It is difficult to speak with certainty on this point, but there can, I think, be little doubt that in many cases the vasomotor and trophic alterations are the result of a lesion (inflammation) in the peripheral nerves, rather than in the spinal cord. This is certainly

the case in the very interesting condition which is termed perforating ulcer of the foot. In fig. 27 I have represented the condition of the tibial and plantar nerves in a case of perforating ulcer of the foot. In that case, the patient suffered from diabetes mellitus; but very similar changes have been found in locomotor ataxia.

The herpetic eruptions are probably also due to inflammatory changes in the peripheral nerves or in the posterior root ganglia; for in herpes zoster, inflammatory changes have been found in these parts.

The exact cause of the osseous lesions is not yet definitely known. As I have already stated, Dr. Buzzard supposes that they are due to a lesion in the medulla oblongata. But the correctness of this theory is, I think, very doubtful. The fact that very similar bone and joint lesions are frequently developed in syringomyelia is highly suggestive that the osseous and joint lesions of locomotor ataxia are due to a lesion in the central grey matter or in the grey matter of the posterior cornu of the spinal cord.

I have now described all the leading symptoms of this most interesting and important disease. In order to complete the clinical picture, I must say a word or two with regard to the general state of nutrition and the complications which may arise in the course of the disease.

General state of nutrition.—The general state of nutrition is in most cases well maintained, for the internal organs are usually healthy and the digestive functions carried on satisfactorily, notwithstanding the constipation which in some cases is very marked. In those cases in which lightning pains, gastric or other visceral crises recur with great frequency and severity, the patient may become thin, careworn and anxious-looking. But even in these cases the patients are often well nourished. In the later stages of the disease, emaciation and marasmus are frequently developed.

Complications.—In the later stages of the disease, the lesion may extend to the anterior cornua or to the crossed pyramidal tract in the lateral column. In some cases, subacute myelitis is developed. Although the sensory cranial nerves or their central connections are not unfrequently affected, the higher cerebral mechanisms are very rarely deranged. Well-marked mental symptoms are very rare, except in those cases in which the disease is associated with general paralysis of the insane (a very

common combination), or in which the patient becomes depressed, low-spirited and melancholic. The mental depression, which intelligent patients, who realise the hopelessness of their condition and who know that the lightning pains which entail so much suffering will probably continue throughout the rest of their lives, suffer from, is often very great; it may be so extreme as actually to suggest the desirability of suicide.

In some cases in which locomotor ataxia and general paralysis are associated, the general paralysis is first developed; in others, the locomotor ataxia precedes the general paralysis; and in others the two diseases, which are due to the same morbid process affecting different parts of the nervous system, are developed simultaneously.

Well-marked syphilitic lesions, such as gummata and softenings due to thrombosis and syphilitic endarteritis of the cerebral vessels, are rarely developed in the course of locomotor ataxia. This is somewhat remarkable, since such a large proportion of the patients have suffered from previous syphilis.

In a certain proportion of cases of locomotor ataxia, valvular or other lesions of the heart occur. I have seen several cases in which the aortic valves were diseased; it is probable that in some of these cases the aortic regurgitation was due to syphilitic disease at the base of the aorta.

In the later stages of the disease, phthisis or other pulmonary complications are apt to be developed.

In those cases in which the bladder is seriously affected (markedly paralysed), cystitis, pyelitis and surgical kidney are of frequent occurrence, and they are often the immediate cause of death.

Types and Course.—Let us now look for a moment at the different types of the disease, and at its course.

As I have more than once told you, the course of the disease is usually very chronic; it is by no means uncommon to meet with cases in which the disease has lasted for twenty or even thirty years.

In typical cases, the disease develops insidiously and progresses slowly, the different stages being distinctly marked and separable from one another. The duration of the different stages varies very considerably in different cases. As a general rule,

the first (pre-ataxic) stage lasts for several, it may be many, months; the second stage (ataxia with ability to walk) for several, it may be for many, years; and the third stage (complete ataxia with inability to walk) also for several years.

In some cases, the onset is more rapid, and the stage of marked ataxia is reached within a few months from the commencement of the symptoms.

Further, we have seen that in exceptional cases the ataxic symptoms seem to be developed at the very commencement of the attack; and that in others, which are by no means uncommon, the disease does not advance beyond the pre-ataxic stage. Cases of pre-ataxic tabes are of great interest. I have seen more than one case in which the pre-ataxic stage lasted for more than twenty years.

In some of the cases of pre-ataxic tabes, the disease is characterised by severe and frequently recurring attacks of lightning pains, loss of the knee-jerks, the Argyll Robertson condition of the pupil, loss of sexual power, constipation, and some impairment of the bladder reflex. In others, some one or other of the rarer symptoms (optic atrophy, gastric or other crises, or the characteristic joint affection), or all of these symptoms are also present.

In rare instances, there are no lightning pains. In one case which I have already brought before your notice, the disease was characterised by frequently recurring gastric crises, loss of the knee-jerks, the Argyll Robertson condition of the pupil, and complete loss of sexual desire and sexual power. In another case, which was probably a case of pre-ataxic tabes, optic atrophy and loss of the knee-jerks were the only symptoms.

Diagnosis.—In typical and fully developed cases, the diagnosis presents no difficulty. The only conditions which a skilled observer is likely to confound with locomotor ataxia, in its fully developed (ataxic) stage, are:—Functional or, as it is usually termed, hysterical ataxia; and the ataxic forms of peripheral neuritis.

In the early (pre-ataxic) stage of the disease, and especially in the rare forms of pre-ataxic tabes in which the lightning pains are slight or absent, the diagnosis is attended with greater difficulty.

The symptoms and signs which are of most importance from

a diagnostic point of view are :—The loss of the knee-jerks ; the lightning pains ; the Argyll Robertson condition of the pupil ; the inability of the patient to maintain his equilibrium when the eyes are closed ; the characteristic difficulty in walking, due, you will remember, not to loss of power but to ataxia ; the impairment or loss of the muscular sense ; and the derangements of the vesical, rectal and sexual reflexes. The loss of the knee-jerks and the Argyll Robertson condition of the pupil are of special value since they are objective signs rather than symptoms, and since they are developed in the early stages of the disease. Loss of the knee-jerk is more important than loss of the pupil reflex to light, for it is almost invariably present ; whereas the Argyll Robertson condition of the pupil is not unfrequently absent.

The differential diagnosis of pre-ataxic tabes and of rheumatism and neuralgia.—In its early or pre-ataxic stage, locomotor ataxia may be confounded with rheumatism or neuralgia (sciatica, for example). This mistake in diagnosis, which used frequently to be made in my student days when our knowledge of locomotor ataxia and diseases of the spinal cord was much less perfect than it is now, is easily avoided, provided that a reasonable amount of clinical care and caution are exercised.

The diagnosis turns upon (1) the character of the pains, and (2) the presence or absence of other symptoms and signs indicative of tabes.

The character of the pains.—The pains of locomotor ataxia are essentially nerve pains ; they occur in paroxysms ; they are usually limited to the legs and lower part of the trunk (pelvic organs) ; they are felt in both legs ; they are rarely limited to the area of distribution of any particular nerve ; they may occur at one part of the leg at one moment, and at another part of the same or the opposite leg or in different parts of the same or both legs within a brief period of time ; they are not attended (or at least very exceptionally attended) by tenderness of the nerve trunks, joints, or muscles, though they may be accompanied or followed by local tenderness of the skin (hyperæsthesia). These characteristics sufficiently distinguish the neuralgic pains of locomotor ataxia from rheumatic pains and the nerve pains of ordinary neuralgia.

Other symptoms and signs of tabes.—But further, in rheuma-

tism and neuralgia, the other symptoms and signs of locomotor ataxia are conspicuous by their absence; whereas, in pre-ataxic tabes, loss of the knee-jerks is almost invariable, and the Argyll Robertson condition of the pupil and some derangement of the vesical, rectal and sexual reflexes are very generally present. No doubt in very rare cases of pre-ataxic tabes, the lightning pains may for a time exist alone (without the loss of the knee-jerks, etc.); but such cases are so exceptional, so infinitely rare, that they need not be taken into practical account.

In doubtful cases (i.e. when the foregoing considerations are insufficient to allow of a positive diagnosis), the age and sex of the patient and the presence or absence of a history of syphilis, may throw light upon the nature of the case.

In the extremely rare cases of pre-ataxic tabes in which some of the rare symptoms (optic atrophy, gastric or other crises, or the peculiar joint lesions) are developed without lightning pains, the diagnosis has to be made in the same way, viz., by observing (1) the exact characters of the optic atrophy, of the visceral disturbance (crisis), or of the joint affection; and (2) the presence or absence of the other symptoms and signs of early tabes (loss of the knee-jerks, the Argyll Robertson condition of the pupil, derangements of the vesical, rectal and sexual reflexes, etc.); and (3) the age and sex of the patient and the presence or absence of a history of previous syphilis.

I need not say that in all cases of suspected tabes, the patient's power of balancing the body and of co-ordinating the movements both of the lower and upper extremities, when the eyes are closed, and the condition of the muscular sense should be carefully tested. In the pre-ataxic stage (i.e. before the ataxia is at all marked) slight impairment of co-ordination can often be detected by delicate tests. Further, in the rare forms of pre-ataxic tabes, in which the lightning pains are inconspicuous and are not spontaneously complained of by the patient, the occasional occurrences of the characteristic pains can often be elicited by cross-examination and direct questioning.

The differential diagnosis of locomotor ataxia with visceral crises and various forms of visceral disease, such as ulcer of the stomach, renal colic, hepatic colic, etc.—In some cases of this kind, an error in diagnosis is easily made; for the visceral symptoms, so to speak, may be very similar in the two cases and the diagnosis is

rendered more difficult by the fact that the visceral crises are usually developed in the pre-ataxic stage of tabes, before the characteristic inco-ordination is developed.

The differential diagnosis must be based upon :—

(1) The mode of onset, duration, and the mode of subsidence of the attack.

(2) The condition of the affected organ between the paroxysms (attacks); and

(3) The presence of other symptoms and signs of pre-ataxic tabes, especially the absence of the knee-jerks, the presence of the Argyll Robertson condition of the pupils, the presence or history of lightning pains, of temporary paralysis of the ocular muscles and of derangements of the vesical, rectal, or sexual reflexes.

Let us take, as an illustration, **the differential diagnosis of the gastric crises of locomotor ataxia and of ulcer of the stomach.**

In locomotor ataxia, the gastric symptoms (pain, vomiting, etc.) usually come on quite suddenly without any apparent exciting (stomach) cause. The appetite and digestive functions are usually quite normal up to the very commencement of the attack. The paroxysms of pain and vomiting are usually more persistent and of longer duration than the paroxysms of pain and vomiting due to an ulcer of the stomach. There is very rarely vomiting of blood; and if blood is vomited it is only in small quantity. The vomited matters consist chiefly of glairy mucus. The patient is usually a male. There is in most cases (if the fact can be ascertained) a history of syphilis.

The attack usually subsides as suddenly as it commenced; and between the attacks (paroxysms) of pain the patient, so far as the stomach is concerned, is usually perfectly well. Immediately after the pain subsides, he may eat freely and without subsequently suffering any pain or other dyspeptic disturbances.

In addition, some of the associated symptoms and signs of pre-ataxic tabes (loss of the knee-jerks, the Argyll Robertson condition of the pupil, etc.) are almost invariably present.

I repeat that the sudden mode of onset without any apparent local causes, in the midst of apparent health so far as the affected viscus is concerned; the severity, long duration and rapid subsidence of the paroxysm; the healthy condition of the suspected viscus between the paroxysms; together with the associated symptoms and signs of pre-ataxic tabes, are the facts

on which the differential diagnosis must also be based in the case of the other visceral crises.

The differential diagnosis of locomotor ataxia and of the various forms of paraplegia.—I have said that, once the ataxia is well developed, the diagnosis is usually unattended with difficulty. It is only an ignorant or careless observer, for example, who could possibly confound locomotor ataxia with paraplegia the result of myelitis.

In locomotor ataxia, the difficulty in walking is due not to loss of motor power, but to inco-ordination; the knee-jerks are absent; and the muscles are sufficiently well-developed. Paraplegia the result of ordinary (transverse) myelitis never presents such a combination of conditions. Further, in cases of locomotor ataxia some of the other characteristic symptoms (lightning pains, Argyll Robertson pupil, etc.) will almost certainly be present.

The differential diagnosis of locomotor ataxia and ataxic paraplegia (postero-lateral sclerosis).—Ataxic paraplegia is distinguished from locomotor ataxia by the exaggerated condition of the knee-jerks, and other points of difference which I need not now detail. You will appreciate them better after I have described the clinical features of ataxic paraplegia.

The differential diagnosis of locomotor ataxia and cerebellar tumour.—Tumours of the cerebellum often produce an ataxic gait; but the diagnosis presents no difficulty. The points of distinction are:—(1) The characters of the ataxia and of the gait; and (2) the nature of the associated symptoms—the presence, in the one case (cerebellar tumour), of symptoms and signs indicative of a coarse intracranial lesion; and, in the other (locomotor ataxia), of the other symptoms and signs indicative of tabes.

The characters of the ataxia and of the gait.—In cerebellar disease, the ataxia is due to the head, not to the heels. A patient affected with cerebellar ataxia reels from side to side like a drunken man; he cannot walk forwards in a straight line; he does not keep the knees straight; he does not throw out the legs in a jerky, sudden and ataxic way, and does not bring the heels down with a stamp. In cerebellar disease, the ataxia affects the muscles of the trunk more than the muscles of the legs; it is often associated with and apparently due to giddiness (vertigo).

The associated symptoms and signs.—When the ataxia is due

to a cerebellar tumour, symptoms and signs of a 'coarse' intracranial lesion (such as headache, vomiting, optic neuritis or post-neuritic atrophy, vertigo, etc.) are always present; and symptoms and signs of locomotor ataxia (lightning pains, loss of the knee-jerks, the Argyll Robertson condition of the pupil, etc.) are conspicuous by their absence.

In cerebellar tumour, the knee-jerks are usually increased and often unequal on the two sides; whereas in locomotor ataxia the knee-jerks are almost always absent on both sides. The condition of the knee-jerks is therefore a point of great diagnostic importance, but it is not always conclusive; for in some cases of cerebellar tumour the knee-jerks, instead of being increased, are abolished. I have seen two or three cases of this kind.

From these statements you will see that there is no real difficulty in distinguishing cerebellar tumour with ataxia from tabes dorsalis with ataxia.

The differential diagnosis of locomotor ataxia and cerebro-spinal sclerosis.—In some cases of disseminated (cerebro-spinal) sclerosis, the gait is markedly ataxic and there is optic atrophy and defective vision; but in typical and well-marked cases of locomotor ataxia on the one hand, and of cerebro-spinal sclerosis on the other, the diagnosis does not present any difficulty.

In typical cases of cerebro-spinal sclerosis, the ataxia resembles that of cerebellar disease rather than that of tabes dorsalis. It is, so to speak, coarser and more widely diffused than the ataxia of tabes dorsalis. In many cases it seems to affect the muscles of the trunk more than the muscles of the lower extremities; and it often affects the muscles of the arms more than those of the legs.

Again, in typical cases of cerebro-spinal sclerosis the knee-jerks are exaggerated and lightning pains and the Argyll Robertson pupil are absent; whereas in locomotor ataxia, the knee-jerks are absent and lightning pains and the Argyll Robertson pupil are usually present. Further, cerebro-spinal sclerosis is attended with other characteristic symptoms, viz., the rhythmical voluntary tremor, vertigo, scanning speech, nystagmus, etc.

These statements apply to typical cases. In quite exceptional cases of cerebro-spinal sclerosis, the knee-jerks are abolished instead of exaggerated; and in very rare cases the Argyll Robertson condition of the pupil is present.

Again, it is possible that in the early stage of cerebro-spinal sclerosis, the lesion might be limited or chiefly limited to the postero-external columns of the spinal cord. In such cases, the diagnosis would be very difficult or impossible, for the symptoms might be identical with, or at all events very similar to, those of locomotor ataxia. But this difficulty in diagnosis is theoretical rather than practical. Such a limitation of the lesion is so extremely rare that it need hardly be taken into practical account.

In cases in which the foregoing points of distinction are insufficient to allow of a positive diagnosis between locomotor ataxia and cerebro-spinal sclerosis (but, as you will have gathered, this is of very rare occurrence) the age and sex of the patient and the presence or absence of a history of syphilis may help to show the true nature of the case.

LECTURE XX

LOCOMOTOR ATAXIA (*Continued*)

At the end of the last lecture, Gentlemen, we were discussing the differential diagnosis of locomotor ataxia; and before passing on to the prognosis and treatment, I must direct your attention to the manner in which locomotor ataxia is to be distinguished from those cases of peripheral neuritis and of functional or hysterical derangement in which ataxia is a prominent symptom.

The differential diagnosis of locomotor ataxia and the ataxic forms of peripheral neuritis.—In some cases of peripheral neuritis (but you must remember that they are rare), the motor symptoms are ataxic rather than paralytic; and since in these cases the ataxia may closely resemble that due to tabes dorsalis, and since the knee-jerks are absent and there are marked sensory derangements (pains, hyperæsthesia, anæsthesia) in the lower extremities, the differential diagnosis may be attended with some difficulty. But the difficulty in diagnosis is not nearly so great as this statement seems to imply. A competent observer, provided that all the facts are taken into consideration, will rarely have any doubt as to the nature of the case. Nevertheless, the similarity of the conditions may be so considerable that I must consider the question of diagnosis in some detail. The chief points of differential distinction are as follows:—

1. *The age and sex of the patient.*—Locomotor ataxia is essentially a disease of the adult male; the ataxic form of neuritis probably occurs as frequently in the female as in the male; the alcoholic form perhaps more frequently in the female than in the male (for alcoholic neuritis is more common in females than in males); the diphtheritic form may occur, and probably usually does occur, at an earlier age than locomotor ataxia.

2. *The rapidity of onset.*—The onset of locomotor ataxia is

usually slow, the onset of peripheral neuritis (due to alcohol, diphtheria, etc.) is usually subacute or rapid.

3. *The previous history and apparent exciting cause.*—Locomotor ataxia usually develops without any apparent (immediate) exciting cause, though in a large proportion of the cases the patient has previously suffered from constitutional syphilis. Peripheral neuritis (especially the alcoholic form) is not of course uncommon in persons who have had syphilis, but a syphilitic history can only be elicited in a small proportion of cases. It is accidental rather than causative. In peripheral neuritis an immediate exciting cause (alcoholic excess, diphtheria, sore throat, etc.) can usually be elicited. I need not go into details nor refer to the difficulty that there sometimes is in eliciting a history of alcoholic excess in the alcoholic cases. These and many of the other points of distinction will be better understood after we have considered the important subject of peripheral neuritis.

4. *The condition of the motor nerve apparatus.*—This and the condition of the vesical, rectal and sexual reflexes are the most important points of distinction. Although in the ataxic forms of peripheral neuritis the motor symptoms are ataxic rather than paralytic, there is usually distinct evidences of paralysis. In the alcoholic form, some loss of power in the extensors of the foot can usually be detected and there may of course be marked foot-drop or wrist-drop. In the diphtheritic form, there is often, or there recently has been, paralysis of the palate or of the ciliary muscle (accommodation). Even in the ataxic forms of peripheral neuritis, there is usually some muscular atrophy, some loss of faradic irritability, and in alcoholic cases more especially, some muscular tenderness on pressure;¹ whereas in locomotor ataxia all of these conditions (paralysis, muscular atrophy, loss of faradic irritability and muscular tenderness) are absent. I am speaking of course of typical cases.

5. *The condition of the pupil, vesical, rectal and sexual reflexes.*—In peripheral neuritis, derangements of the pupil, bladder, rectum and sexual apparatuses, which are so characteristic of locomotor ataxia, do not occur or only accidentally occur.

6. *The exact character of the sensory disturbances.*—In peripheral neuritis, the pains do not as a rule present the paroxysmal

¹ Tenderness of the calf muscles (when pinched) is highly characteristic of the alcoholic form of peripheral neuritis.

lightning character of locomotor ataxia; they are usually more localised to particular nerve areas; they are often associated with tenderness of the nerve trunks, and, in the alcoholic form, with muscular pain and tenderness. In the alcoholic cases in particular, there is often intense objective hyperæsthesia of the skin of the soles. When, for instance, a blunt instrument such as the sharp end of a tuning-fork is sharply drawn across the sole, the patient often screams out from the intense suffering (burning sensation) which results.

7. *The mental condition of the patient.*—In peripheral neuritis, the mental condition is often characteristically affected. But I need not go into details, for the ‘ambulatory delusions’ as I term them are rarely (if ever) present in the ataxic form of the disease, and this is of course the only form which is likely to be confounded with locomotor ataxia.

8. *The course and progress of the case.*—In doubtful cases, the progress of the disease and the effects of treatment will clear up the diagnosis. The course of locomotor ataxia is to progress from bad to worse or at the best to remain stationary. The course of peripheral neuritis is to get well. Recovery is generally slow, often very slow, but (unless there are complications) it is usually sure.

The differential diagnosis of locomotor ataxia and of functional ataxia.—Dr. Charlton Bastian has directed attention to the fact that extreme ataxia, with loss of the muscular sense and, it may be, with profound disturbances of sensation (anæsthesia, analgesia, etc.) in the ataxic or other parts of the body, occasionally occurs.

These cases are very rare, but they are of great scientific interest. They are also of practical importance, for in some of their features they closely resemble locomotor ataxia—a disease which is so unamenable to treatment; whereas they (the functional cases) almost invariably in the course of time get well.

There is of course no difficulty in discriminating the ordinary form of so-called ‘hysterical ataxia’ in which the patient walks with a jerky and irregular gait and in which the defect is chiefly psychical in character. In such cases the difficulty in walking is more a difficulty depending *on idea* than on true ataxia. In the cases to which I am referring, the ataxia is real. It may be absolutely indistinguishable from the most extreme ataxia of tabes dorsalis. The muscular sense may be entirely lost. So

long as the patient is able to regulate (govern and direct) the movements of the affected limb or limbs with the aid of sight, the movements may be performed with some degree of accuracy, but when the eyes are closed she (for these cases rarely occur in men) is unable to make any co-ordinate movement. Standing or walking when the eyes are closed may, when the lower extremities are affected, be absolutely impossible. Indeed, in extreme cases, the patient may be unable when the eyes are closed to move the limb at all; and when it is (passively) moved, to realise how it was moved or to say where it is situated (to find the foot, etc.).

A case to which I have already more than once directed your attention is illustrative of this difficulty in diagnosis. It further shows the chief facts to which attention should be directed in order to arrive at a correct opinion. They are as follows:—

1. *The age and sex of the patient.*—Functional ataxia is much more common in women than in men, and in young than in middle-aged women.

2. *The rapidity of onset and apparent cause.*—The onset of functional ataxia is usually more rapid than that of tabes dorsalis; indeed in some cases the ataxic symptoms are quickly developed after an accident, injury, emotional disturbance, illness, prolonged course of nursing, etc. The functional form of ataxia is not a result of syphilis, although in rare cases an accidental history of syphilis may of course be forthcoming.

3. *The associated symptoms.*—This is the most important point. In functional ataxia, lightning pains, loss of the knee-jerks and the Argyll Robertson condition of the pupil are, so far as I know, never present, and any derangement of the bladder which may be present is usually temporary.

Loss of the knee-jerks is a most important point of distinction, for in the functional forms of ataxia (which are probably due to a cerebral rather than a spinal derangement) the knee-jerks are usually increased. It remains for future observation to show whether there may not be a functional form of ataxia due to disturbance of the spinal cord in which the muscular sense and the knee-jerks are both lost. If this variety of functional ataxia does occur, it could only be distinguished from locomotor ataxia by the age and sex of the patient, the rapidity of

onset, the absence of previous syphilis, the absence of lightning pains and the Argyll Robertson condition of the pupil, and the presence of associated symptoms indicative of hysteria.

Further, the functional form of ataxia is often associated with other symptoms and signs of hysteria (well-marked hemianæsthesia, ovarian tenderness, emotional manifestations, hysterical fits, etc.). The presence of other hysterical manifestations does not, of course, conclusively show that the ataxia is functional; for, as I have more than once pointed out, hysteria is often associated with, and the result of, organic disease. But the presence of other hysterical manifestations is highly suggestive that the ataxia is functional.

In the case which is related on page 316 the diagnosis was very difficult. The patient was a young woman; there was no reason whatever to suppose that she had had syphilis; the ataxic and paraplegic symptoms developed rapidly after a definite exciting cause (prolonged nursing, mental anxiety and exhaustion); the patient was undoubtedly emotional (hysterical); lightning pains were very slight; there was the most extreme ataxia and complete loss of the muscular sense in both lower extremities; the knee-jerks were abolished; the Argyll Robertson condition of the pupil was present; and the functions of the bladder were in some (a slight) degree affected. Further, there was no improvement under treatment.

When all of the facts of the case were taken into consideration (and especially the loss of the knee-jerks, the Argyll Robertson condition of the pupil, and the absence of any improvement under treatment), there could be no doubt that the ataxia was the result of organic disease, and the subsequent progress of the case has shown that this opinion was correct.

Prognosis.—In most cases of locomotor ataxia, the prognosis as regards arrest and cure is very unfavourable, but fairly good as regards the duration of the disease. In the majority of cases, the disease pursues a progressive course; the lesion is sometimes arrested, and in rare and exceptional cases the symptoms entirely disappear and a cure seems to be established. In most of the cases in which a complete cure takes place the disease is probably pseudo-tabes.

In forming a prognosis, the individual circumstances and

special features of each case must, of course, be taken into account. In some cases, the disease runs an unusually rapid course; in others, more especially the pre-ataxic cases in which optic atrophy is developed, the course is very prolonged; but it must not be forgotten that some of the visceral crises, especially laryngeal crises, which are usually developed in these pre-ataxic cases, are not unattended with danger. In those cases in which cystitis, pyelitis, aortic disease, phthisis and other grave complications are developed, the prognosis is, of course, much more serious. Fortunately, the prognosis is not always so unfavourable in respect to arrest and cure as the statement which I have just made would imply; for in a not inconsiderable proportion of cases, temporary periods of improvement and it may be of apparent arrest occur.

Since syphilis is such an important factor in the production of the disease, one might perhaps, *a priori*, expect that locomotor ataxia would be beneficially influenced by antisymphilitic treatment, and that in those cases in which the disease develops soon after the primary sore, the prognosis would be more favourable than in the average run of cases. In some cases this is perhaps the fact; but such a fortunate result is in my experience very rare. Speaking generally, we may I think say that, in the present state of our therapeutic knowledge, the syphilitic element is of little or no importance for the purposes of prognosis and treatment. We have seen that many of the symptoms of locomotor ataxia may be produced, not only by a sclerotic lesion in the posterior columns of the cord, but by a lesion in the peripheral nerves. It is probable, as I have already pointed out, that in many of the rare cases in which the symptoms clear up and disappear under iodide of potassium and mercury, the lesion involves the peripheral nerves rather than the spinal cord. But be this as it may, the existence of previous syphilis does not materially benefit the prognosis in the great majority of cases.

In not a few cases of locomotor ataxia, the progress of the lesion seems to be arrested after it has advanced up to a certain point. But arrest is not cure; for, in cases of this kind, the lightning pains, the ataxia and the other symptoms (which may be present) usually continue.

The prognosis as regards duration and non-development of

further symptoms is most hopeful in those cases in which the pre-ataxic stage is of long duration. In some cases, as I have more than once told you, the ataxic stage is never reached. This is a most important consideration, so far as the patient is concerned; for, so long as there is no ataxia, he can move about and engage in various occupations which, if he were ataxic, he could not follow. You must not, however, forget that in many of these cases of pre-ataxic tabes the lightning pains are of terrible severity, and that the repeated occurrence of severe lightning pains knocks the patient, as it were, out of time, and leaves him in a prostrate and debilitated condition.

Further, in those cases in which visceral crises, optic atrophy or joint lesions are developed, the prognosis (so far as the ability of the patient to follow an occupation or profession is concerned) is obviously worse than in the cases in which lightning pains are the only or the chief symptoms.

In short, in this as in almost every disease, the prognosis must be guided by the special features of each individual case.

Optic atrophy usually progresses, once it is developed, and may ultimately lead to the production of complete blindness. In very rare cases, the loss of sight improves under treatment. I have already referred to one case of this kind which has come under my own notice.

In trying to form an opinion as to the severity of the disease in any given individual case, we have to take into account:—The length of time which the disease has lasted; the rate at which it seems to be progressing; the exact character of the symptoms; the presence or absence of complications; the effects of treatment; and the age and circumstances of the patient. After the exhaustive way in which we have considered the clinical history of the disease, you will readily appreciate the prognostic significance of the different symptoms. I need not go into details; but by way of illustration I may say that the early development of a marked degree of ataxia is very unfavourable; that paralysis of the bladder and especially of the sphincter muscle is a serious symptom; and that such complications as free aortic regurgitation, kidney disease or phthisis are of very grave significance.

Treatment.—Let us now turn to the treatment of the disease. We have seen that although syphilis plays a most important

part in the production of locomotor ataxia, it is only in a few rare and exceptional cases that antisyphilitic remedies exert any beneficial effect upon the progress of the disease. That, at all events, is the opinion of the great majority of physicians. It entirely represents my own experience. Personally, I have rarely, if ever, seen any decided benefit result from iodide of potassium and mercury, even in cases in which the disease has developed soon after the primary sore. I remember one case in particular in which the symptoms of locomotor ataxia developed three years after the initial lesion and in which no benefit whatever resulted from antisyphilitic treatment. But although this is the result of my own experience, I am not prepared to say that iodide of potassium and mercury are always useless. In the hands of some physicians these remedies seem to have been beneficial, and occasionally, it is said, curative. Now, since the disease cannot be cured or indeed materially influenced for good by any known plan of treatment, it is only right to give the patient the benefit of the chance, and to see whether in his particular case the antisyphilitic plan of treatment will prove useful. Possibly some of us have failed to obtain beneficial results with iodide of potassium and mercury in cases of locomotor ataxia because we have not given these remedies a sufficiently long and vigorous trial. But it is well to remember that a vigorous antisyphilitic treatment, when it fails to do good, is apt to do harm. It may be stated as a general rule, that, on the one hand, in those cases in which iodide of potassium is beneficial, it is well borne; and on the other, that in those cases in which it fails to do good, it is often badly borne. In some forms of tertiary syphilis, enormous doses of iodide of potassium are not merely tolerated but are obviously beneficial. Patients will sometimes tell you that they feel that every dose of the medicine does them good. They seem to suck in the iodide and to thrive on it, if I may use the simile, as they would do on their mother's milk. On the other hand, some persons are so constituted that a single grain of iodide of potassium is poisonous to them; many healthy persons bear iodide of potassium very badly. In some cases of locomotor ataxia, iodide of potassium produces marked depression and is undoubtedly harmful. The remedy must not therefore be blindly prescribed, and the effects which it produces in each individual case must be closely watched.

For the reasons which I have just stated, it is, I think, advisable to commence the treatment of a case of locomotor ataxia by giving iodide of potassium. A small dose, say five grains of the iodide three times a day, may be first prescribed. If the remedy is well borne, the dose should be gradually but rapidly increased up to twenty or thirty grains three times daily. If at the end of three or four weeks (that is to say, after a fair trial) there is no apparent improvement, or if the iodide seems to disagree, the remedy should be discontinued altogether.

An essential point in the treatment of locomotor ataxia is to see that the patient avoids everything which is likely to aggravate the symptoms and to hasten the development and progress of the lesion. The general health must, so far as possible, be kept in the highest state of efficiency; the patient should be well housed, well clothed, and well fed; the condition of the digestive organs must be carefully attended to; this is especially important in those cases in which there are gastric crises; constipation, which in some cases is very obstinate, should be carefully corrected. If the patient can afford it, he should pass the winter and spring months in a warm equable climate (Egypt, Algiers, Tangier, etc.), for exposure to cold and damp are certainly prejudicial; while abundance of sunlight and fresh air are beneficial. If the patient is obliged to remain in this country during the inclement winter and spring months, he should, so far as possible, avoid exposure to cold and damp. A certain amount of muscular exercise is beneficial; but muscular effort which tires and exhausts and produces overfatigue is bad. All depressing conditions should, so far as possible, be avoided. Mental anxiety, worry, and anything which causes mental depression undoubtedly have a prejudicial effect upon this, as indeed they have upon every other chronic nervous disease. But in many cases it is, of course, impossible to avoid mental anxiety and worry. Patients who are not well off, especially married men with wives and families dependent on them, must go on working as long as possible. If their occupation is a sedentary and indoor one, they are fortunately in many instances able to follow it for years even after the ataxia is considerable. A late distinguished Professor in the University held one of the most important medical Chairs for several years after the disease had reached the second stage. I know several patients affected with

locomotor ataxia who are leading useful lives. They are of course heavily handicapped, not only by the ataxia but by the recurring attacks of lightning pains. In cases of this kind, the relief of the pains is a most important part of the treatment. I will return to this point presently.

Excesses of all kinds, especially excesses which throw a strain upon the spinal cord, must be avoided. In this, as in every form of nervous disease, alcoholic excess is very prejudicial. The excessive use of tobacco is also no doubt injurious; but smoking in moderation may in my opinion be allowed. To many of us tobacco is a sedative, a solace and a great source of enjoyment. Those of you who are smokers will understand what a comfort the pipe is likely to be to many locomotor patients. In some cases, it is one of the few sources of enjoyment which remains to them; and as I have already said, there are, so far as I know, no grounds whatsoever for supposing that the moderate use of tobacco is injurious.¹ Of course in every case in which there is reason to suppose that optic atrophy is commencing, the use of tobacco should be absolutely prohibited.

Sexual excess is very harmful; but except in rare cases it is only in the early stages of the disease that the patient requires to be cautioned on this point; in most cases of locomotor ataxia, the sexual appetite is impaired or lost, when the patient comes under the care of the physician.

It is hardly necessary to warn ataxic patients against the risk of accidents and falls; they know by their own experience the danger of attempting to walk in crowded thoroughfares or on slippery pavements, etc.

Quinine, the mineral acids, the compound syrup of the phosphates, cod-liver oil, and similar remedies are useful as general tonics; but arsenic, nitrate of silver and strychnine are (excluding iodide of potassium and mercury, to which I have already referred) perhaps the only drugs which exert any directly beneficial effect upon the disease. I am satisfied myself that arsenic and strychnine in particular are in many cases beneficial. Many other remedies (ergot, belladonna, phosphorus, etc.) have been recommended; but I am doubtful if any one of them has any special influence upon the disease.

¹ 'Smoking does not injuriously affect the busy man who thinks; it only hurts the lazy man who drinks.'

Electricity has of course been used in this as in every other nervous malady. The statements which I have previously made with regard to the direct application of the constant current to the spine in cases of primary spastic paraplegia apply equally here. I very much doubt whether a weak constant current applied to the spine can do any good; but it can do no harm, and since many people believe that the catalytic action of the constant current has a beneficial effect upon the sclerotic process, I see no objection to trying it, provided that it is cautiously and judiciously used.

In those cases of locomotor ataxia in which the muscles are firm and well nourished, the local application of the faradic current is unnecessary. In those cases in which the muscles are soft and flabby or wasted, faradic stimulation applied directly to the muscles is a most important and useful means of promoting and keeping up the muscular nutrition. I may perhaps take this opportunity of telling you that the faradic current has also been employed for the relief of the lightning pains; in some cases it seems to do good; in others it completely fails.

Counter irritation over the affected part of the cord is, I think, useful in some cases, especially in those cases in which there is pain in the back (due to, or suggestive of, associated meningitis), and in which the disease is rapidly (acutely) developed. In cases of this kind, the actual cautery should be freely applied with the aid of an anæsthetic. If the patient does not benefit from the first application, the cauterisation need not be repeated.

Hydropathic measures (bathing and douching of various kinds), which are strongly recommended by some German writers, are seldom employed in this country; perhaps we undervalue hydropathy and are not sufficiently acquainted with the effects which it produces.

Nerve-stretching was fashionable a few years ago, but has now fallen into well-merited disuse. Doctors, like some women, are perhaps too apt to follow the whim of the moment whatever it may happen to be. Like crinolines and other peculiarities of dress, which are anything but an adornment to the female form divine, methods of treatment, which more extended experience shows to be of little value and which are sometimes even the reverse of beneficial, are apt for a brief period to become the fashion.

Experience has shown that, although nerve-stretching does in

some cases relieve the lightning pains and produce perhaps a temporary improvement in the ataxic symptoms, it is powerless to arrest the progress of the disease and that it is not unattended with danger; it may even be followed by death itself. No one, so far as I know, now thinks of recommending nerve-stretching, in this country at all events.

The suspension plan of treatment followed nerve-stretching. Very different results were obtained by different observers. It seems certain that some cases are for a time materially benefited by the treatment; but most authorities are, I think, now agreed that the effects are, at the best, palliative (temporary relief of pain or ataxia); that in many cases there is no beneficial effect at all; and that the treatment is not absolutely free from danger. Suspension is contra-indicated in cases in which the heart, arteries, or lungs are diseased, in very fat and heavy patients, and in cases in which laryngeal crises occur.

The most recent method of treating locomotor ataxia which has been introduced is the subcutaneous injection of (a) animal extracts (extracts of spinal cord, brain, nerve and testicle); and (b) of a solution of phosphate of soda. Extraordinary results have, it is said, been obtained by the phosphate of soda method. It will be remarkable if these results are corroborated by the experience of the profession.

The relief of the lightning pains is one of the most important points in the treatment of locomotor ataxia. Various remedies have been employed for this purpose. Morphia, phenacetin, antipyrin, antifebrin, salicylate of soda, exalgine and Indian hemp are the most useful drugs. It is well to defer the use of morphia as long as possible, and to try, in the first place at all events, whether relief cannot be obtained by some of the other remedies which I have just named. Phenacetin is in my experience (leaving morphia out of account) by far the most reliable drug; but antipyrin, antifebrin and exalgine in some cases do remarkably well. To be effective, the phenacetin must be given in full doses. I am in the habit of prescribing twenty grains at the commencement of the attack, and, if the first or second dose does not give relief, repeating the same dose every hour until sixty grains have been taken. The effects of the remedy must be carefully watched. In my experience unpleasant effects are very rarely produced even by these large doses.

Painting the skin with a strong solution of cocaine, the subcutaneous injection of cocaine at the seat of the pain, painting the skin with a 20% solution of menthol in olive oil, rubbing a small quantity of aconitia ointment over the seat of local pain (care being taken that the surface is unbroken), and the application of the faradic brush to the surface of the skin, are the most important local remedies. They are all very uncertain in their effects; but they undoubtedly give relief in some cases in which the pain and hyperæsthesia are localised and superficial.

In many cases, the only means of relieving the sufferings of the patient is the hypodermic injection of morphia; but as I have already told you, the administration of morphia should be deferred as long as possible, lest the morphia habit should be established. The patient should be earnestly cautioned not to administer the morphia, either by the mouth or subcutaneously, to himself. Once he begins to dose himself with morphia, he will be apt to carry the practice to excess. Dreading the terrible pain, and knowing the happy relief which morphia gives him, he will be only too apt to take the drug on the slightest provocation. Whenever he feels a slight pain, he will, thinking a paroxysm is coming on, take a dose of morphia. It is quite a question whether the morphia habit is not a worse complaint than the original disease. I would advise you, therefore, to thoroughly exhaust every possible plan of treatment before you prescribe the continued and systematic use of morphia. But in many cases, the lightning pains are so severe that the patient must have relief. If you do not relieve him he will take to dosing himself. It is well to recognise this, and, when other means fail, to have recourse to morphia; for it is eminently desirable that the morphia treatment, which in many cases is the only certain means of relief, should be regulated by the physician rather than by the patient.

For relief of the gastric crises, the same remedies which I have recommended for the relief of the neuralgic pains (morphia, phenacetin, antipyrin, antifebrin, etc.) are to be employed. Hydrocyanic acid, carbolic acid, menthol (dissolved in spirits of chloroform or aromatic spirits of ammonia) are in some cases useful. Blisters applied to the epigastric region and along the course of the vagi nerves in the neck may also be tried. In

some cases, the application of electricity over the course of the vagi nerves in the neck seems to be attended with benefit.

The other forms of visceral crises should be treated on the same principle, viz., the administration of internal remedies and local applications which allay pain, spasm and irritation.

When the bladder is paralysed, special care must be taken to prevent the development of septic cystitis and the kidney complications which are apt to result from it. I shall return to this point in more detail when I come to describe the treatment of myelitis.

During the later stages of the disease (i.e. after the patient has become bedridden), the prevention of bed-sores requires attention.

In those cases in which optic atrophy is developed, iodide of potassium or mercury or both may be tried, but strychnine, given by the mouth or subcutaneously, is probably the best remedy.

The tendency to the production of trophic lesions must be remembered; all sources of local irritation should be avoided: painful corns should be soothed and softened rather than cut; blisters on the soles, ulcers and sores about the toes or on any other part of the body should be carefully attended to.

LECTURE XXI

FRIEDREICH'S ATAXIA

TO-DAY, Gentlemen, I propose to direct your attention to Friedreich's ataxia, a disease which in some of its features closely resembles locomotor ataxia, but which is a definite and distinct affection.

Synonyms.—The disease was first described by Friedreich in the year 1861 as a variety of locomotor ataxia. The synonyms '*Hereditary ataxia*,' and '*The congenital form of locomotor ataxia*' have been applied to it. Dr. Ladame of Geneva and the late Professor Charcot proposed to call the disease *Friedreich's disease*. They objected to the terms 'the congenital form of locomotor ataxia' and 'hereditary ataxia,' because the ordinary typical form of locomotor ataxia is sometimes, though very rarely, hereditary; and because Friedreich's disease is not so much a hereditary as a family disease, that is to say, it is not usually handed down directly from father to son through succeeding generations, but it attacks several members of the same generation. I shall return to this point presently.

For my own part I think the term *Friedreich's ataxia* is preferable to Friedreich's disease. As a general rule it is not desirable to name diseases after their discoverers. Such terms as Friedreich's disease, Graves' disease, Kaposi's disease convey no information as to the nature of the disease which they are used to designate, at least to one who is unacquainted with these affections. In naming a disease, it is desirable to adopt some term which describes concisely the nature of the disease, or which gives some information as to the nature of the lesion on which the disease depends. Hence, the term Friedreich's ataxia is, I think, much better than the term Friedreich's disease. Such terms as mitral stenosis, poliomyelitis anterior acuta, multiple cerebro-spinal sclerosis are admirable, for they suggest some

of the leading features of the diseases to which they refer. But in selecting such terms it is necessary that they should be correct, at all events as far as they go. If the name given to a disease implies a false pathology, it is even more objectionable than a term which implies no pathology at all. Now, if Friedreich's ataxia is a distinct disease and not a mere variety of locomotor ataxia (ordinary tabes), the term 'the hereditary form of locomotor ataxia' should not be given to it.

Definition or Short Description.—Friedreich's ataxia is a chronic disease of the spinal cord (? and medulla oblongata), which usually begins insidiously in childhood or about the age of puberty (almost always before the age of twenty), and which slowly but steadily progresses in a downward direction until it ultimately terminates in death.

It is a rare disease which seems almost as common in girls as in boys, and which usually affects several members of a family.

In typical and fully developed cases, it is characterised by:—Inco-ordination which involves the arms as well as the legs; an ataxic-cerebellar gait; loss of the knee-jerks; thickness of speech; a form of club-foot; curvature of the spine; a vacant heavy expression of countenance; and choreic-like twitchings and movements.

Etiology.—As I have already told you, the disease is essentially a family affection. Several members of the same family (brothers and sisters) or of the same generation (cousins) are usually affected.

A remarkable series of cases occurred in a family called Blattner. The great-great-great-grandfather of the present generation, who married in the year 1710, seems to have been affected with the disease, for he was nick-named 'the stumbler.' Rutimeyer, who reports the cases, states that all of the direct descendants of this original Blattner (the great-great-great-grandfather of the cases of Friedreich's disease which he describes) are inscribed under the surname of stumbler (*Stulzie*) in the Official Register of the Commune of Kuttigen in the year 1840; and he remarks that the gait of this man must have made a great impression upon his contemporaries, since it remained for so many generations in the memory of the inhabitants of the

country. Four families who are the direct descendants in the male line of this original Blattner (the 'stumbler') exist at the present time. In all of these families some cases of Friedreich's disease have appeared, and yet it is very remarkable that no case of the disease has occurred in the intermediate generations.

So far as we at present know, isolated cases are rare.

In this tendency to attack several members of the same family or generation, Friedreich's ataxia resembles pseudo-hypertrophic paralysis and Thomson's disease. Although very few isolated cases have been recorded, it is, I think, exceedingly doubtful whether they are so rare and exceptional as is usually supposed. It is, I think, not unlikely that the isolated cases often pass unrecognised. When several members of a family are affected, the diagnosis is not only very much more easy, but the disease impresses itself as being something peculiar on the minds both of the parents and the medical attendant.

It necessarily happens that Friedreich's disease can very seldom be directly handed on from parent to child, for the disease being one of youth is in most cases already far advanced before the patient reaches the marriageable age. The same statement applies to pseudo-hypertrophic paralysis. In the great majority of cases the subjects of Friedreich's disease and pseudo-hypertrophic paralysis are obviously quite unfit to marry when they reach the age of puberty. This inability to marry is in no way due to non-development of the sexual organs, for the sexual development takes place as usual at the ordinary time of puberty, but to the fact that the patients when they attain the marriageable age are usually more or less helpless and crippled. As a matter of fact, however, several cases have been reported in which patients affected with Friedreich's ataxia have married and had children. In some instances, the ages of the children born under such circumstances are perhaps hardly as yet sufficiently advanced to say confidently that they will not be affected with the disease; but in others, it must, I think, be concluded that the children have escaped. Whether the disease will again manifest itself in their children, as it did in the celebrated Blattner family, remains of course to be seen.

So far as I know, there is only one case on record in which it is absolutely certain that both parent and son were affected with the disease.

In this tendency to affect several members of the same family, Friedreich's ataxia presents a very remarkable contrast to ordinary tabes.

The relatives (fathers, mothers, uncles, aunts, grand-parents, etc.) of patients affected with Friedreich's ataxia have, in many instances, been insane or suffered from some other form of nervous disease; in other words, the subjects of Friedreich's ataxia often inherit a tendency to nerve disease of some kind or another, though seldom to Friedreich's ataxia itself.

In many instances, the families affected with Friedreich's ataxia have been remarkable for their numbers. It must also be noted that in several of these families a number of still-births have occurred.

In several of the recorded cases, the parents or other near relatives of patients affected with the disease have been notably intemperate; and some writers have suggested that conception when one or other of the parents was intoxicated is a cause of the disease. But the correctness of this theory is very doubtful.

In a considerable proportion of the recorded cases of Friedreich's ataxia, the disease seems to have developed after recovery from an acute febrile disease (measles, scarlet fever, small-pox, etc.). It would consequently appear that in persons, who, by inheritance or by some family peculiarity, are predisposed to Friedreich's ataxia, an acute febrile disease may act as an exciting cause of the condition.

Dr. Newton Pitt has advanced the theory that the reason why the disease is so frequently developed after an acute febrile affection and at or about the time of puberty, is in consequence of the vascular alterations which occur as a result of these conditions; he believes that the cord lesion is perhaps due to endarteritis.

Syphilis does not seem to play any part in the production of Friedreich's ataxia. In this respect the disease presents a very remarkable contrast to locomotor ataxia. In that disease, as I have already pointed out, syphilis is an etiological factor of the highest importance; in the few cases of genuine locomotor ataxia which have been met with in children, the patients always seem to have inherited syphilis. It is of course quite possible that in a true case of Friedreich's ataxia a history of inherited syphilis may now and again be forthcoming. But this is clearly an accidental circumstance, for in almost every case of the disease

which has so far been recorded, it is definitely and distinctly stated that syphilis could be excluded.

Members of the same family are usually attacked by the disease at or about the same age. This is of some importance for prognosis; for the children of a family (affected with Friedreich's ataxia) who pass the critical age without the disease developing will probably escape.

Morbid Anatomy.—In Friedreich's ataxia, as in the ordinary form of tabes, the posterior columns of the cord are sclerosed; but the lesion is not confined to the posterior columns; the lateral columns are also affected.

The cases of Friedreich's ataxia which have been examined post mortem are not numerous. Some of them occurred many years ago, when the different tracts of the spinal cord were very imperfectly differentiated, and when the methods of histological research were very much less complete than they are now. In others, the disease was of very long duration; in cases of this kind—and they comprise almost all the cases which have been examined post mortem—it is difficult to form a correct opinion as to the exact nature and position of the primary and fundamental lesion.

Recent observations seem to show that the spinal cord is distinctly smaller than normal; and that, in some cases, this congenital defect (for it must be considered a congenital defect) involves the medulla oblongata and the pons Varolii as well as the spinal cord.

The lesion seems to be a complicated one—a sclerosis which involves (*a*) the columns of Goll; (*b*) the columns of Burdach; and (*c*) the crossed pyramidal tracts. In some cases (*d*) the vesicular columns of Clarke; and (*e*) the direct cerebellar tracts are also affected (see figs. 114 to 118).

In some cases—but this is probably the result of the extension of the primary lesion—other parts in the lateral and anterior columns of the cord are affected, viz., Gowers' tract, the so-called mixed ascending tract in the lateral column, and the direct pyramidal tracts in the anterior column.

From this statement you will see that the cord lesion in Friedreich's ataxia is much more extensive than the cord lesion in ordinary tabes.

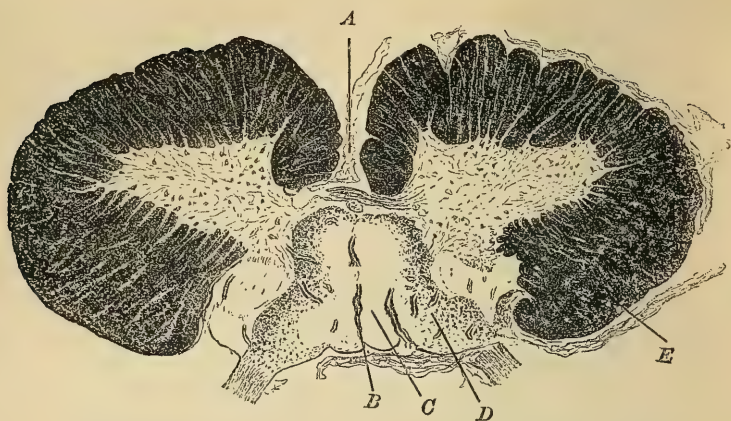


FIG. 114.—*Transverse section of the spinal cord in the cervical region stained by Weigert's method.*—(After Déjerine and Letulle.)

A. Anterior median fissure: the anterior parts of the antero-lateral column are absolutely normal. B. Posterior median fissure. C. Column of Goll, unstained, in consequence of a high degree of sclerosis. D. Fasciculus of Burdach, partly sclerosed. E. The posterior part of antero-lateral column, slightly paler than normal, corresponding to the direct cerebellar tract.

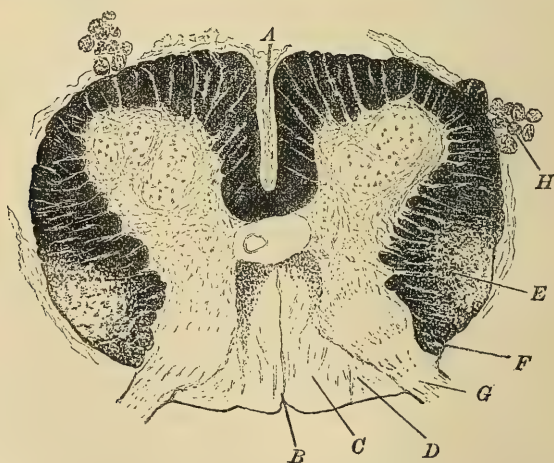
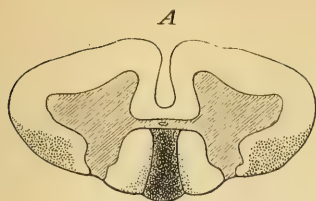


FIG. 115.—*Section through the lumbar region of the same cord.*

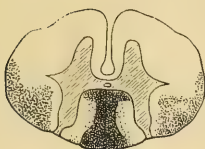
The sclerosis of the posterior columns is much more marked than in the cervical region.

The letters have the same significance as in the preceding figure. E points to the antero-lateral column; the sclerosis in this part of the cord is limited to the crossed pyramidal and direct cerebellar tracts.

FIG. 116.



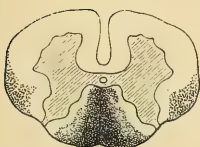
B



C



D



E

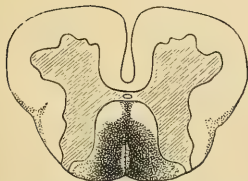
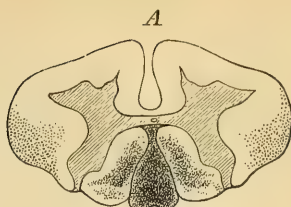
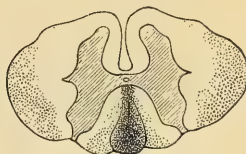


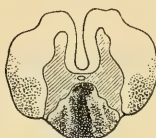
FIG. 117.



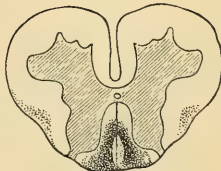
B



C



D



E



FIGS. 116 and 117.—A series of sections through the spinal cord in Rüttimeyer's two cases of *Friedreich's ataxia*.—In these cases the chief characteristic lesions were:—a high degree of degeneration of the posterior columns, more especially of Goll's columns, throughout their whole extent, and a systematic degeneration of the crossed pyramidal tracts and of the direct cerebellar tracts. There was no affection of the direct pyramidal tracts. In the grey matter, Clarke's columns and their cells were degenerated. The anterior horns were unaffected. Lissauer's tracts were not involved. The posterior roots were degenerated.

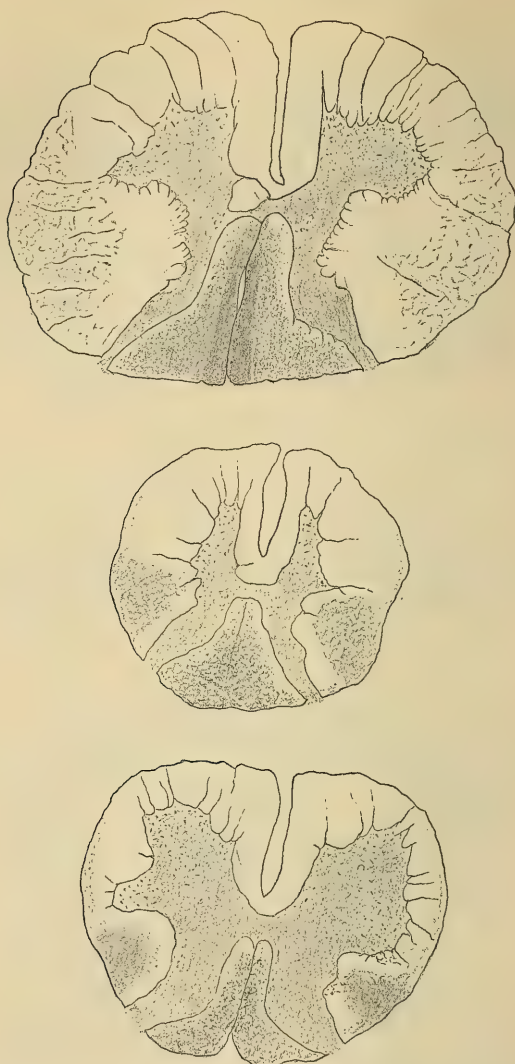


FIG. 118.—Transverse sections through the cervical, dorsal, and lumbar regions of the spinal cord in a case of *Friedreich's ataxia* (reported in the author's '*Atlas of Clinical Medicine*,' vol. i. page 46—Case 2).

The postero-external as well as the postero-internal columns are sclerosed in the cervical, dorsal and lumbar regions.

In the cervical region there is some scattered sclerosis in the lateral columns, more especially in the situation of the crossed pyramidal tracts.

In the dorsal and lumbar regions the crossed pyramidal tracts are markedly sclerosed.

Again, the lesion in the posterior columns of the cord does not appear to be identical in the two affections. In ordinary tabes, the sclerosis of the postero-external columns is generally confined to the lower parts of the cord; but in Friedreich's ataxia the postero-external columns are always more or less sclerosed in the cervical as well as in the dorsal and lumbar regions. Further, it is said that Lissauer's tract, which is always markedly sclerosed in ordinary tabes, is not usually involved in Friedreich's ataxia.

Again, in typical cases of ordinary tabes the posterior roots are profoundly degenerated and the peripheral nerves are often markedly affected; while in Friedreich's ataxia the posterior roots are, comparatively speaking, little degenerated and the peripheral nerves are either not involved at all or only in a very slight degree.

It must also be noted that the fine sensory nerves in the muscles, which are perhaps always affected in the ordinary form of locomotor ataxia, are not, so far as we at present know, involved in Friedreich's ataxia.

Opinions differ as to the exact significance of the lesion of the crossed pyramidal and direct cerebellar tracts. Most authorities regard the sclerosis in these tracts as primary and systematic. According to this view, the cord lesion in Friedreich's ataxia is a combined system lesion of the columns of Goll and the columns of Burdach in the posterior columns, and of the crossed pyramidal and direct cerebellar tracts in the lateral columns of the spinal cord.

But Déjerine and Letulle take a different view. They think that the sclerosis of the postero-external and postero-internal columns is primary and systematic, but that the sclerosis of the crossed pyramidal and direct cerebellar tracts is secondary and non-systematic. They further think that the sclerosis in the lateral columns presents all the characteristic features of a sclerosis due to a cortical meningo-myelitis. But in opposition to this latter statement it must be remembered that in some cases of Friedreich's ataxia it is expressly stated that the membranes were not adherent, thickened or opaque.

Déjerine and Letulle are of opinion that the sclerosis of the posterior columns in Friedreich's ataxia is essentially different from that of ordinary locomotor ataxia, and indeed from every

other sclerotic lesion which has hitherto been described. They look upon the sclerosis of the posterior columns in Friedreich's disease as a pure neuroglial sclerosis. 'The appearance,' they say, 'of the posterior columns is quite different from that which one see in ordinary tabes. In transverse sections, the fibres are

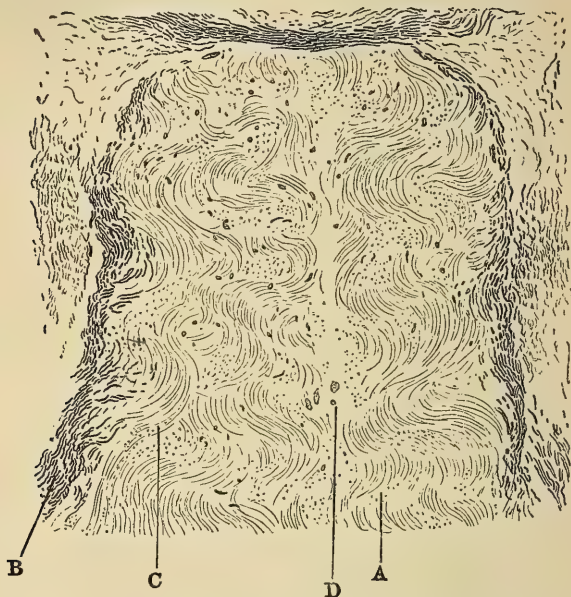


FIG. 119.—*Transverse section through the posterior columns at the level of the lumbar enlargement.*—(After Déjerine and Letulle.)

The sclerosis is seen to consist of whorls of neuroglial fibres crowded together. The vessels are sufficiently numerous.

A points to very long fasciculi running parallel with the plane of the section; the serpentine undulations of the fibrils give the section a very remarkable appearance.

B points to a reticulated appearance, due to the transverse divisions of fasciculi of neuroglial fibres which are placed perpendicularly to the surface of the section.

C points to some nerve fibres still surrounded by their myeline sheath.

D. The posterior median fissure showing a transversely divided vessel filled with blood.

seen to cross one another in different directions, and to form true whorls exactly like those which M. Chaslin has described in cases of epilepsy. (See figs. 119 and 120.) The nerve tubes, instead of appearing transversely cut, are seen not unfrequently running in a direction longitudinal to the surface of the section. In other words, they follow the direction of the whorls in which they

happen to be situated. This is a peculiarity which has not been previously pointed out in Friedreich's ataxia, and which is always absent in ordinary tabes.' They add: 'Under all the methods of preparation and staining the same appearances are seen, and

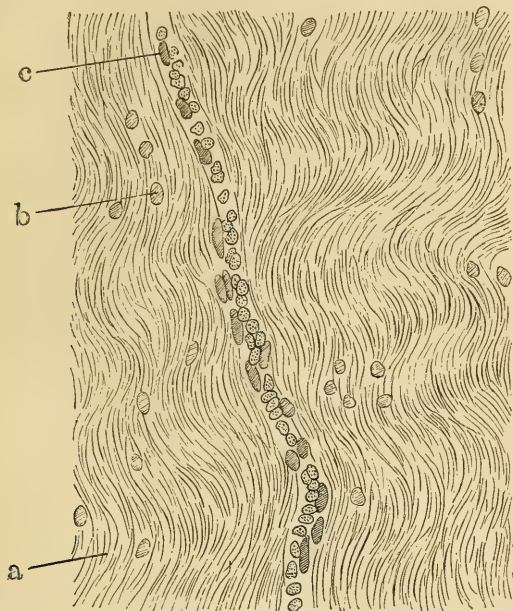


FIG. 120.—The appearance of the neuroglial fibres more highly magnified; from a longitudinal section of the posterior columns at the level of the lumbar region.—(After Déjerine and Letulle.)

- a. Neuroglial fibrils lying parallel to the section.
- b. Nuclei of neuroglial corpuscles.
- c. Nuclei of the endothelial cells lining a healthy capillary blood-vessel passing through the neuroglial tissue. The blood corpuscles which it contains are misshaped in consequence of the method of preparation (Malassez's method, viz. carmine, potash, acetic acid).

we are able to affirm with certainty that the process with which we are dealing does not present any analogy with the vascular sclerosis with which we are acquainted, and that the condition is a pure neuroglial sclerosis.'

Clinical History.—As I have already stated, the disease usually commences in early life. The symptoms may be described as partly positive and partly negative; for the recognition of the

disease, as a definite and distinct clinical entity, is based partly on the presence of certain symptoms (the *positive* symptoms), and partly upon the absence of others which are characteristic of some of the other diseases with which Friedreich's ataxia is most likely to be confounded (the *negative* symptoms).

The more important positive symptoms are:—Ataxia; impairment of the motor power of the legs, and especially of the muscles on the front of the legs; a peculiar gait; absence of the knee-jerks; deformity of the foot; lateral curvature of the spine; thickness of speech; nystagmus; choreic-like twitchings; inability to maintain the erect position when the eyes are closed; and in some cases unprovoked and uncontrollable attacks of laughter, palpitation, occipital headache and vertigo.

The ataxia first involves the lower extremities and the trunk, then passes upwards and affects the upper extremities, the tongue, eyeballs, etc.

The ataxia does not seem to be a sensory ataxia, properly so-called.

The sensibility of the skin, the muscular sense and the special senses are usually quite normal, at all events until the later periods of the case. This is an important point of distinction between Friedreich's ataxia and ordinary tabes.

In the absence of any obvious sensory impairment it is difficult to suppose that the ataxia in Friedreich's disease is due (as it appears to be chiefly due in the ordinary form of locomotor ataxia) to interruption of ingoing impressions.

It is probable, I think, that the ataxia in Friedreich's disease is due to interruption of the commissural connections which pass from segment to segment through the posterior columns; in other words, to the same derangement of the spinal co-ordinating mechanism which is probably in part at least the cause of the ataxia in the ordinary form of locomotor ataxia.

I need not go into details, for I have already considered the question (see page 295).

Loss of motor power in the legs is not usually present in any marked degree, at all events in the earlier stages of the disease; but in the later stages, paralysis with contractures or muscular atrophy is frequently developed.

In some of its features, *the gait* of Friedreich's ataxia resembles that of locomotor ataxia; in others, that of cerebellar disease. It has consequently been termed *ataxic-cerebellar*.

A patient affected with locomotor ataxia walks in a straight line; but a patient affected with Friedreich's ataxia staggers from side to side like a drunken man. In Friedreich's ataxia, the feet are not usually raised so high and the heels are not brought down with the characteristic thump of locomotor ataxia. In well-marked cases the feet are often inverted as the patient walks.

Again, in Friedreich's ataxia, the gait is much more shaky (the head and trunk as well as the legs shake) than in locomotor ataxia. The patients often tumble or strike themselves against articles of furniture. Indeed, in the advanced stages of the disease the unsteadiness is so marked that one dreads to let the patient walk alone. In many cases, the head is poked forward when the patient walks, or rather when he staggers forward in an irregular and uncertain manner. In some cases, the patient looks as if he were being forcibly propelled forwards; his walk is half a run; he staggers heavily forward with his head down, looking as if he were trying to overtake his centre of gravity in front of him. This festinating gait, as it has been termed, is not developed in all cases of the disease.

In consequence of the ataxia in the upper extremities, the patient finds it difficult to button his clothes, and to perform other delicate movements. In many cases, when the patient attempts to pick up a small object, such as a pin, the hand is suddenly jerked forwards. Charcot has likened this action to that of a bird swooping down upon its prey.

Inability to maintain the erect position when the eyes are shut and the feet placed close together (Romberg's symptom), which is invariably present in fully developed cases of ordinary tabes, does not seem to be a constant feature of Friedreich's ataxia. The statements of different observers, however, differ on this point.

Absence of the knee-jerks is a highly characteristic symptom of the disease and is of great diagnostic importance as distinguishing Friedreich's ataxia and insular (disseminated) sclerosis. Loss of the knee-jerks is an early, probably the earliest, symptom; it may, in fact, be present before any other features of the disease are developed. And it is curious to note that in some cases the knee-jerks have been absent in the healthy (non-ataxic) members of ataxic families, i.e. in the healthy brothers and sisters of patients affected with the disease.

Speech affection.—Speech is almost always affected in Friedreich's ataxia. The alteration resembles more or less closely that of insular sclerosis. The voice is thick, low-toned, monotonous,

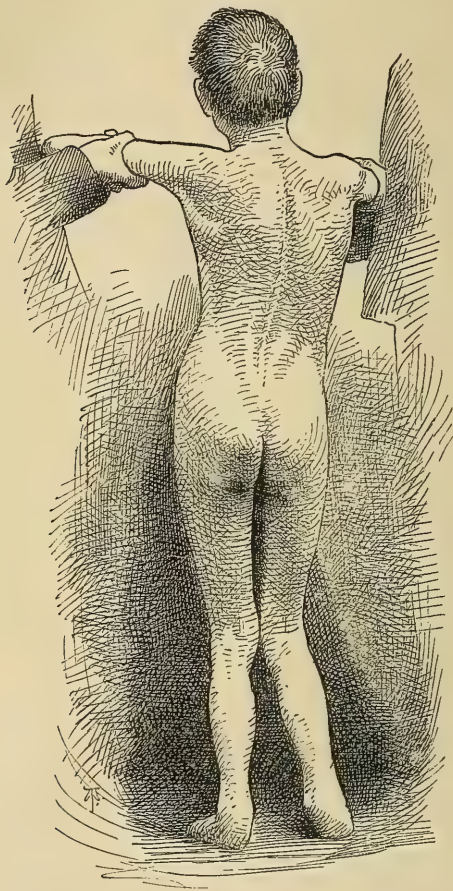


FIG. 121.—Case of *Friedreich's ataxia* seen from behind.

There is a well-marked curvature of the spinal column towards the right; this is not sufficiently brought out in the drawing; the right side of the back should project much more than it does in the figure.

hoarse or harsh, and the articulation slow and apparently difficult. In many cases, the patient speaks as if he had a foreign body in the mouth, or as if he were the worse for

liquor. The staggering, reeling gait, the thick speech and the vacant somewhat stupid expression, which is present in many cases of the disease, are all at first sight suggestive of drunkenness. In one of my cases the patient was actually accosted in



FIG. 122.—*The patient represented in fig. 121 two years later; the spinal curvature is much more marked.*

the middle of the day by a policeman who thought he was drunk. As a rule, the speech affection does not become well developed until the other symptoms of the disease are well marked.

The vacant expression of countenance to which I have just referred, resembles more or less closely that which is seen in some cases of chorea and in cerebro-spinal sclerosis. In many cases this vacant expression is not developed until the later periods of the case.

Choreic-like movements are very generally present. They occur when the muscles are at rest, and usually involve the

muscles of the leg and trunk; less frequently those of the face, tongue and upper extremities. They are often developed in the early stages of the disease, and in some cases seem to precede the ataxia. Cases of Friedreich's ataxia are consequently in their early stages not unfrequently mistaken for cases of ordinary chorea.



FIG. 123.—*The foot in a case of Friedreich's ataxia.*

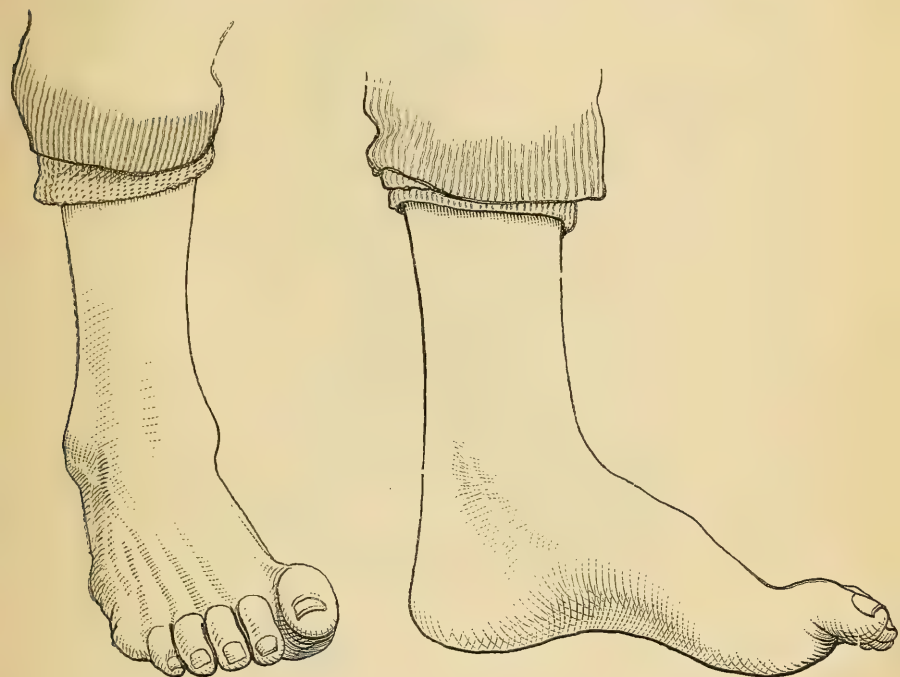
In most well-marked cases of Friedreich's ataxia, *curvature of the spinal column* is developed (see figs. 121 and 122). In some cases, the curvature is in a lateral, in others in an antero-posterior direction; and not unfrequently the two forms of curvature are combined.

The foot deformity which I have already incidentally mentioned is a striking and characteristic feature of the disease (see figs. 123, 124 and 125). It is best seen when the patient

is sitting and the foot is raised from the ground. The foot looks short and stumpy; it appears to be compressed from before backwards in the antero-posterior diameter; the instep is highly arched; and the dorsum of the foot very prominent. The balls of the toes, especially of the great toe, appear to be enlarged. The first phalanges of the toes, particularly that of the great toe, are over-extended on the metacarpal bones, and the extensor

FIG. 124.

FIG. 125.



FIGS. 124 and 125.—*Feet in a case of Friedrich's ataxia. Standing position.*

tendons, especially the extensor proprius pollicis, stand prominently out. In many cases, the feet are cold and of a red or purple colour. When the patient is standing and leaning his weight upon the foot, the deformity is much less marked, the shortening, the arching of the instep and projection of the dorsum are then to a large extent effaced. (See figs. 124 and 125.) But even in the standing position the extension of the first phalanx and the prominence of the extensor tendons can

usually be seen. This deformity of the foot is in many cases developed at a comparatively early stage of the disease, and is, therefore, of considerable diagnostic significance. It is not, however, pathognomonic; in more than one case of pseudo-hypertrophic paralysis I have seen a very similar condition.

The exact cause of the foot deformity is not definitely known. It seems to be a form of paralytic club-foot, and is by some



FIG. 126.—*Friedreich's ataxia in an advanced stage (Dr. Everett Smith's case).*

authorities thought to be due to the lesion of the crossed pyramidal tract in the lateral column of the cord.

It is interesting to note that the right foot of the father of one of my cases of Friedreich's ataxia presented the characteristic foot deformity, without exhibiting any other indications of the disease.

Nystagmus is usually a late symptom, and sometimes difficult to detect. In order to elicit it, the patient should be made to direct the eyes as far in a lateral direction as possible, or to fix an object placed just in front of the nose.

In many of the recorded cases of the disease, *unprovoked and uncontrollable attacks of laughter* have occurred. In others, *palpitation of the heart* has been observed. In several of the reported cases *valvular lesions* have been present. Déjerine and Letulle suggest that the mitral stenosis which was present in their case was probably congenital.

The general health is usually unimpaired; but in the later stages of the case, when the patient is reduced to the position of a helpless cripple, the general health not unfrequently becomes affected.

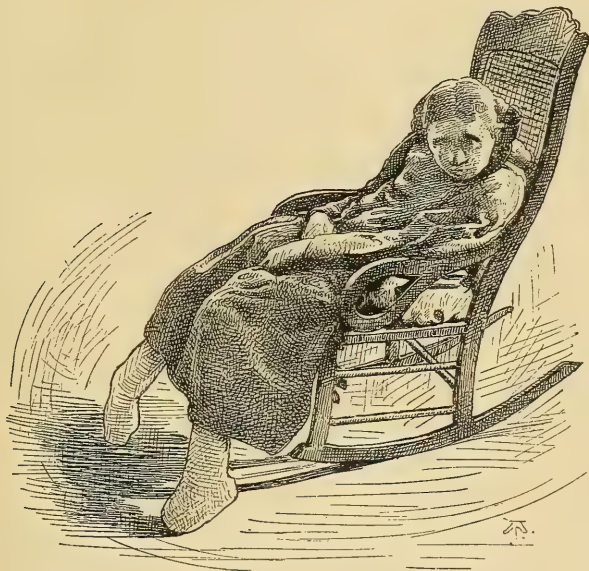


FIG. 127.—*Friedreich's ataxia in an advanced stage (Dr. Everett Smith's case).*

Negative symptoms.—The more important negative symptoms and signs are as follows:—

1. Absence of lightning pains, and of any marked impairment of the muscular sense, of the sensibility of the skin to touch, heat, cold, pain, and of the special senses (sight, hearing, taste and smell).

Lightning pains have been met with in a few cases. In the later stages of the disease, a considerable degree of anæsthesia of the skin is not uncommon. In exceptional cases, the cutaneous

sensibility and the muscular sense appear to be impaired in the earlier stages of the disease. Hyperæsthesia of the soles is present in some cases, and perhaps accounts for the fact that the plantar reflex is sometimes exaggerated.

2. Absence of any derangement of the bladder and rectum, and of the sexual apparatus.

3. Absence of the Argyll Robertson condition of the pupil, of myosis, optic atrophy and temporary paralysis of the external muscles of the eyeball.

4. Absence of trophic derangements of the bones, joints, skin, hair, nails, teeth, etc.

5. Absence of headache, vomiting, optic neuritis, congestive or pseudo-apoplectic attacks, and derangements of the memory and intellectual faculties.

Vertigo has been met with in a considerable number of cases. Occipital headache has been occasionally noted; in the advanced stages of the disease epileptiform attacks occasionally occur.

Insanity has been recorded in one instance; but in the great majority of cases of the disease, cerebral symptoms (if we exclude the affections of speech, the nystagmus and vertigo) are conspicuous by their absence.

Course and duration.—The course of the disease is usually slow; but, as I have more than once stated, the lesion tends to progress steadily from bad to worse, until finally the patient is reduced (provided he lives long enough) to the condition of a helpless cripple. (See figs. 126 and 127.)

LECTURE XXII

FRIEDREICH'S ATAXIA (*Continued*)

IN the last lecture, Gentlemen, we considered the clinical history of Friedreich's ataxia. Let us now turn to the diagnosis.

Diagnosis.—The disease is, as we have seen, a distinct clinical and pathological entity. The diagnosis is chiefly difficult when it occurs in an isolated form, i.e. when only one member of a family is affected. Friedreich's ataxia is one of the rarest of all known forms of nerve disease. The great majority of medical men have never had an opportunity of seeing it; they are consequently unacquainted with its characters. Further, one does not expect to meet with it in an isolated form. This is one of the chief sources of error in diagnosis.

The recognition of the disease is to be based upon:—(1) the presence of the positive symptoms and signs; and (2) the absence of the negative symptoms and signs, which have been enumerated above.

The diseases with which Friedreich's ataxia is most likely to be confounded are:—Locomotor ataxia, disseminated cerebro-spinal sclerosis, ataxic paraplegia, chorea and cerebellar tumour.

The differential diagnosis of Friedreich's ataxia and of ordinary tabes.—From the description of the clinical history which has been given above, it will be apparent that the two diseases present many points of resemblance. Both are very chronic in their course, both are usually—Friedreich's ataxia always—progressive and incurable. In both, the posterior columns of the cord are sclerosed; in both, difficulty of walking, which is due to inco-ordination and not to paralysis, is the leading symptom, at all events in the earlier stages of the case; in both, the knee-jerks are abolished. But on close analysis it is apparent that the ataxia itself and the gait are not identically the same in the two affections. In

Friedreich's ataxia, the gait is ataxic-cerebellar in character (see the description given above). In Friedreich's ataxia, the inco-ordination almost invariably, in the course of a few years, affects the upper extremities; whereas in ordinary tabes the upper extremities are, comparatively speaking, rarely involved. In Friedreich's ataxia, lightning pains and, as has already been mentioned, derangement of the sensory functions of the skin and of the muscular sense are usually conspicuous by their absence; whereas in ordinary tabes lightning pains and some derangements of the sensory functions of the skin and of the muscular sense are almost invariably present. Eye symptoms, such as the Argyll Robertson condition of the pupil, myosis, optic atrophy and temporary paralysis of some of the external muscles of the eyeball, are very rarely indeed observed in cases of Friedreich's ataxia, but are of frequent occurrence in ordinary tabes. Derangements of the bladder and rectum and of the sexual functions are rarely seen in Friedreich's disease, but are generally present in ordinary tabes. Gastric, renal and intestinal crises, and vasomotor and trophic disturbances (joint disease, bone disease, herpetic eruptions, derangements of the nutrition of the hair, nails, teeth, etc.) are very seldom if ever seen in Friedreich's ataxia, but are met with in a certain proportion of cases of ordinary tabes. Further, the characteristic alteration of speech, spinal curvatures, the foot deformity, choreic twitchings, nystagmus, and the vacant expression of countenance, which characterise Friedreich's ataxia, do not occur in the ordinary form of tabes.

The age of the patient and the fact that several members of the family are in many cases affected with the same disease, are important diagnostic features. True tabes is extremely rare before the age of twenty; and in those cases in which it has been observed in young subjects the patients have almost without exception been the subjects of inherited syphilis. Whereas, Friedreich's ataxia is almost invariably developed in early life, and in patients who are rarely, if ever, the subjects of inherited syphilis. The ordinary form of tabes is very much more common in men than in women. Friedreich's ataxia seems to affect girls almost as frequently as boys.

The differential diagnosis of Friedreich's ataxia and of multiple cerebro-spinal sclerosis.—In both affections, there is difficulty in

walking; in both, speech is involved very much in the same way; and in both, nystagmus and vertigo are often present. The two diseases are, however, absolutely distinct. The distinction is even more radical than between Friedreich's ataxia and ordinary tabes; for the pathological anatomy of Friedreich's ataxia and multiple cerebro-spinal sclerosis is totally different; whereas, the pathological anatomy of Friedreich's ataxia and of ordinary tabes is to this extent the same, that in both diseases the posterior columns of the cord are sclerosed. Friedreich's ataxia and cerebro-spinal sclerosis are distinct clinical and pathological entities; they differ materially in their etiology, symptomatology, clinical history and course; even the gait, the speech and the nystagmus—the three symptoms which they have chiefly in common—are not identical.

The *gait* in Friedreich's ataxia is ataxic-cerebellar; it appears to be a combination of the gait of locomotor ataxia and of cerebellar disease; while the gait of disseminated sclerosis when well marked is either spastic, spastic-cerebellar, or rhythmical, trembling and spastic. In cerebro-spinal sclerosis, the feet are raised with apparent difficulty from the ground; in advanced cases they may appear to be stuck to the ground, and the march is broken up, as it were, by the fine rhythmical movements which are such characteristic features of the disease. In many cases of cerebro-spinal sclerosis, the knee is flexed when the foot is raised from the ground; but in Friedreich's ataxia, as in ordinary tabes, there is a tendency to keep the knee extended.

Although the *speech* of Friedreich's ataxia resembles more or less closely that of multiple sclerosis, the articulation is not, as a rule, so drawled out; the distinction between the different syllables is not so marked—in short, the speech is not so 'syllabic.'

In cerebro-spinal sclerosis, *nystagmus* is usually much more marked and is developed at an earlier period of the disease than in Friedreich's ataxia.

Friedreich's ataxia is, as we have seen, a family disease, and isolated cases are, so far as our present knowledge enables us to judge, comparatively rare. Cerebro-spinal sclerosis, on the other hand, seldom if ever occurs as a family disease; the cases are almost invariably isolated. Friedreich's disease almost always develops before the age of twenty; whereas cerebro-spinal sclerosis usually develops after the age of twenty. Cases of disseminated

sclerosis are, however, sometimes met with in children. In these cases, the diagnosis is more difficult. The onset of Friedreich's disease is usually very slow and gradual and the course is always steadily downwards, from bad to worse. So far as we at present know, periods of remission and distinct improvement seldom, if ever, occur. Whereas, in cerebro-spinal sclerosis, remarkable remissions or periods of temporary improvement are by no means uncommon. In some cases of cerebro-spinal sclerosis, the onset appears to be abrupt; head symptoms, an epileptic fit or a pseudo-apoplectic attack may occur at the onset or in the early stages of the case.

The rhythmical tremor which develops during voluntary movement, but which is completely absent so long as the patient is at rest, is a highly characteristic feature of cerebro-spinal sclerosis. In Friedreich's ataxia, the voluntary movements are rendered unsteady and jerking by the ataxia and by the choreic twitchings which are so often present; but, so far as I know, a true voluntary rhythmical tremor, such as characterises disseminated sclerosis, is never met with in this disease.

In cerebro-spinal sclerosis, the knee-jerks are usually exaggerated, and ankle-clonus is often present; whereas in Friedreich's ataxia the knee-jerks are absent and ankle-clonus is never present. The condition of the knee-jerks is consequently a most important point of distinction between the two diseases.

The foot deformity and the spinal curvature, which are such characteristic features of well-marked cases of Friedreich's ataxia, rarely if ever occur in cerebro-spinal sclerosis.

Cerebral symptoms, vertigo, loss of memory, and pseudo-apoplectic attacks are of frequent occurrence in cerebro-spinal sclerosis, but are rarely met with in Friedreich's ataxia. In fact, some patients affected with Friedreich's ataxia are remarkable for the keenness of their intellectual powers. Atrophy of the optic discs and constriction of the visual fields occur in a considerable proportion of cases of cerebro-spinal sclerosis, but are seldom, if ever, so far as our present knowledge enables us to judge, met with in Friedreich's ataxia.

The differential diagnosis of imperfectly-developed (atypical or *fruste*) cases of cerebro-spinal sclerosis and Friedreich's ataxia may be very difficult; and it is important to remember that in not a few cases of cerebro-spinal sclerosis the characteristic

rhythmical tremor may completely disappear for a time. In cases of this kind, the diagnosis is, of course, particularly difficult. But it is the precocious cases of cerebro-spinal sclerosis—the cases which develop during childhood—that are most liable to be confounded with cases of Friedreich's ataxia. A careful consideration of the clinical features of the two conditions, in particular the condition of the knee-jerks and the presence or absence of the foot deformity, will usually enable the observer to come to a correct conclusion as to the nature of the case.

The differential diagnosis of Friedreich's ataxia and of chorea.—As I have already mentioned above, choreic-like movements are frequently present in cases of Friedreich's ataxia. In those cases in which the choreic-like movements are developed in the earlier stages (i.e. before the other characteristic symptoms are well marked) the disease is easily mistaken for ordinary chorea; but a very little consideration will enable a careful observer to distinguish between the two conditions. In Friedreich's ataxia, the choreic movements chiefly affect the muscles of the legs and trunk, less frequently the face, tongue and upper extremities; whereas, in ordinary chorea, the muscles of the face, tongue and upper extremities are more frequently, or at least quite as severely, affected as those of the lower limbs. Ordinary chorea is usually developed acutely, but the onset in Friedreich's ataxia is slow and gradual. In ordinary chorea, the irregular movements disappear and the disease is cured in the course of a few weeks or months; but in Friedreich's ataxia, even if the choreic movements diminish in intensity or disappear other symptoms persist and develop. The differential diagnosis must, however, be chiefly based upon the presence or absence of the other characteristic symptoms indicative of Friedreich's ataxia. Among these, the absence of the knee-jerks, the ataxia, the peculiar gait, the foot deformity and the characteristic alteration of speech are the most important. When the choreic-like movements are due to Friedreich's ataxia, some of these symptoms will invariably be present; whereas, in ordinary chorea they are all conspicuous by their absence. The family history is, I need not say, a point of great diagnostic importance. Mistakes are only likely to be made in those cases in which the Friedreich's ataxia affects only one member of a family. When several other members of the family are suffering or have suffered from the disease, notwith-

standing the presence of choreic-like movements which may at first sight simulate ordinary chorea, the true nature of the case is obvious enough.

Prognosis and Treatment.—So far as we at present know, the disease invariably tends to progress steadily and continuously in a downward direction until it finally terminates in death. The course is almost always exceedingly slow and chronic. Several cases have been recorded in which the disease has lasted for twenty or thirty years.

In the later stages, the patients are usually reduced to the position of helpless cripples, unable to do anything either with their arms or legs; in some cases speech is so much affected that they are unable to make themselves intelligently understood.

The rapidity with which the disease develops, and the length of time which elapses before the patient is unable to walk, vary considerably. In some of the recorded cases the subjects of Friedreich's ataxia have married and had families. It is needless to say that even in the slighter forms of the disease, marriage must be sternly discouraged, should the physician be consulted on the point.

No remedial measures which have been employed seem to have the least effect in arresting the progress of the lesion. Arsenic, strychnine, nitrate of silver and other remedies, which appear to be beneficial in some cases of locomotor ataxia, may be tried.

The application of the constant electrical current to the spinal column has been recommended by some writers. Suspension, which produces temporary benefit in some cases of locomotor ataxia, does not appear to be of any use in Friedreich's ataxia. A well-fitted plaster jacket, by the support which it affords to the muscles of the trunk, is undoubtedly helpful in some cases; it tends to prevent the development of the spinal curvature which in the later stages is apt to become so conspicuous.

The general health must of course be maintained in the highest possible state of efficiency. The patient should be allowed to walk about as long as possible; but when the ataxia becomes marked they should be cautioned to be careful lest they should injure themselves by falls.

ATAXIC PARAPLEGIA

(COMBINED SCLEROSIS OF THE POSTERIOR COLUMNS AND CROSSED PYRAMIDAL TRACTS).

IN our study of locomotor ataxia, we have seen that the lesion occasionally extends to the lateral columns, and that in Friedreich's ataxia the crossed pyramidal or other tracts in the lateral columns are usually involved in a greater or less degree.

The essential feature of the condition to which I wish to direct your attention is a sclerosis of the posterior columns and crossed pyramidal tracts.

The clinical features represent this combination. The chief symptoms are, (1) ataxia, due to sclerosis of the posterior columns, and (2) muscular weakness and exaggeration of the deep reflexes, due to the lesion of the crossed pyramidal tracts.

The disease is rare; I need not describe it in any great detail. It will be sufficient to indicate its chief characteristics and to direct attention to the points in which it differs from locomotor ataxia and Friedreich's ataxia. But before indicating these differences, let me refer in a little more detail to the morbid anatomy.

Morbid Anatomy.—I have said that the disease is due to a combined sclerosis of the posterior columns and crossed pyramidal tracts, but the sclerosis is in some respects peculiar. In the first place, it differs from the sclerotic lesion of locomotor ataxia inasmuch as the *maximum* lesion (sclerosis) is usually more marked in the dorsal rather than in the lumbar region of the spinal cord. It is possibly owing to this fact that the kneejerks, which are lost in ordinary locomotor ataxia, are retained and exaggerated. In the second place, the sclerotic lesion usually appears to be less intense in the postero-external columns than in typical cases of ordinary locomotor ataxia. This perhaps explains the remarkable clinical circumstance that lightning

pains are almost always absent. The absence of lightning pains is a highly characteristic feature of the disease, and constitutes a notable point of difference between ataxic paraplegia and the ordinary form of locomotor ataxia. In the third place, the lesion in the lateral columns is not usually sharply circumscribed to the pyramidal tracts; it is a much more diffuse lesion than the sclerotic lesion which results from secondary descending degeneration. In this respect it corresponds with the lesion in the lateral column which is met with in Friedreich's ataxia and in primary lateral sclerosis. In some cases of ataxic paraplegia, the direct pyramidal tracts are involved; in others, the ascending cerebellar tracts or the other fibres and tracts of the lateral column are affected.

Further information is required before it is possible to speak definitely with regard to the pathological anatomy. The disease is rare; it is very seldom fatal, and very few post-mortem examinations have been made. Personally, I have had no opportunity of examining any case after death, although I have seen several which presented all the clinical features of the disease during life.

Etiology.—The disease usually attacks adults between 30 and 50 years of age. Men are much more frequently affected than women. The exciting cause is in many cases obscure. Exposure to cold is often thought to be the cause. Sexual excess has also been blamed. Traumatic injury is sometimes said to be an exciting cause. The disease is very rarely preceded by syphilis, and in those cases in which there is a syphilitic history the previous syphilis was probably merely coincident. This is a most important point of difference between ataxic paraplegia and ordinary locomotor ataxia; for, as we have already seen, in the great majority of cases of ordinary locomotor ataxia syphilis is *the* etiological element of chief importance. In the rare cases in which ataxic paraplegia seems to be developed acutely or subacutely, the primary lesion is probably a myelitis, which subsequently progresses and leads to the production of sclerosis both in the posterior and lateral columns. A striking case of this sort, in which the disease was apparently due to the sudden arrest of menstruation, came under my notice a few months ago.

Clinical History.—The onset is usually insidious. The first symptom is generally difficulty in walking, partly the result of unsteadiness and ataxia, partly the result of muscular weakness. When the disease is well developed, a dull pain in the lower part of the back or sacral region is often complained of. In many cases, there is loss of sexual power; in one of my cases the patient was completely impotent. As I have previously remarked, lightning pains, which are so prominent and characteristic of locomotor ataxia, are usually completely absent. In rare cases, the patient complains of a girdle pain. In one of my cases this symptom was very marked.

On examination, the muscles are found to be well-developed, often indeed unusually firm. The knee-jerks are exaggerated, often markedly so, and ankle-clonus can in most cases be obtained.

The ataxia resembles, but is not exactly identical with, that of ordinary locomotor ataxia. Though the patient is unsteady when he walks, the ataxia is especially evident when he attempts to turn. The ataxia in many cases seems to involve the muscles of the back more than those of the lower extremities. The high-action, stamping gait which is so characteristic of locomotor ataxia is rarely if ever marked; further, in ataxic paraplegia there is a greater tendency in walking to sway from side to side and to diverge from a straight line.

Constipation is usually a prominent symptom. There may be some weakness of the detrusor urinæ, but rarely any marked paralysis of the bladder, at all events in the earlier stages of the disease.

In most cases, there are no objective derangements of sensation, though the patient not unfrequently complains of numbness. Romberg's symptom, swaying when the eyes are closed and the feet placed close together, is usually marked. In some cases the muscular sense seems to be considerably impaired. The Argyll Robertson pupil is rarely present; indeed, the ocular derangements which are so conspicuous in many cases of locomotor ataxia are in the great majority of cases of ataxic paraplegia entirely absent; there is little if any tendency to the production of optic atrophy. Joint lesions, vasomotor alterations and visceral crises practically never occur.

As the disease progresses, the weakness and difficulty in

walking and the spastic condition increase. In one case, in which the symptoms were typical and in which the patient has been under my observation for the past twelve years, the condition is now one of complete spastic paraplegia. The patient is unable to stand or walk; the legs are rigidly extended and adducted. Probably this case is exceptional; for, so far as my experience enables me to judge, such an extreme degree of rigidity and tension is rarely developed even in the later stages of the disease.

Gowers states that in some exceptional cases the knee-jerks are lost and that in others the skin sensibility is impaired and lightning pains, optic atrophy or the Argyll Robertson condition of the pupil are developed. No case in which any of these symptoms occurred has come under my own notice, but from the character of the lesion one would naturally expect that lightning pains would occasionally be developed. In short, in some cases of ataxic paraplegia, the clinical features more closely resemble those of locomotor ataxia than of spastic paralysis; whereas in others, the ataxic symptoms are less marked and the spastic symptoms are more prominent.

As in ordinary spastic paraplegia, the lesion may ultimately extend to the anterior cornua, with the production of localised muscular atrophy.

In the great majority of cases the symptoms are for the most part limited to the lower extremities and to the muscles of the back; but it is by no means uncommon to have the upper extremities involved in some degree. It is even stated that in some cases the face muscles are involved.

The general health is, in the great majority of cases, good. In the later stages of the disease, the bladder may become paralysed and kidney complications may be developed.

The course is usually very slow, but in most cases, as in primary lateral sclerosis and locomotor ataxia, the tendency is to progress and to get worse.

Diagnosis.—In typical and well-marked cases, the diagnosis presents little or no difficulty to a physician who is acquainted with the clinical features which the disease presents. The chief diseases which have to be distinguished from ataxic paraplegia are the ordinary form of locomotor ataxia, Friedreich's ataxia and ordinary spastic paraplegia.

The differential diagnosis of ataxic paraplegia and locomotor ataxia.
 —By far the most important diagnostic point is the condition of the knee-jerks—exaggerated in ataxic paraplegia, abolished in locomotor ataxia. But, while this is the general rule, it must be remembered that exceptions to both statements occasionally occur. In rare cases of ataxic paraplegia, the knee-jerks are said to be lost; and in very rare cases of locomotor ataxia the knee-jerks are retained though rarely if ever exaggerated in the earlier stages. Other corroborative points in favour of ataxic paraplegia are:—the absence of a history of syphilis, the absence of lightning pains, of the Argyll Robertson condition of the pupil and of other ocular symptoms, the absence of sensory disturbances, of vasomotor and trophic derangements, and of visceral crises.

The differential diagnosis of ataxic paraplegia and of Friedreich's ataxia.—Here again, the most important point is the condition of the knee-jerks—exaggerated in ataxic paraplegia, lost in Friedreich's ataxia. Corroborative evidence is also afforded by the age of the patient and by the family history. Ataxic paraplegia rarely occurs except in adult males; Friedreich's ataxia is almost invariably developed in childhood or youth. Further, the characteristic speech affection, nystagmus, vacant expression of countenance and foot deformity of Friedreich's ataxia are all absent in ataxic paraplegia.

The differential diagnosis of ataxic paraplegia and of lateral sclerosis, whether primary or secondary.—This rarely presents any difficulty. The essential characteristic is, that in ataxic paraplegia there is muscular inco-ordination as well as loss of muscular power and exaggeration of the reflexes.

The differential diagnosis of ataxic paraplegia and of cerebellar tumour.—In both conditions, the gait may present very similar features, i.e. may be more or less reeling and inco-ordinate; and in both conditions, the knee-jerks may be exaggerated and there may be loss of power in the lower extremities. But in ataxic paraplegia the characteristic symptoms and signs of an intracranial tumour (severe headache, vomiting, optic neuritis or post-neuritic atrophy—all of which are well marked in cases of cerebellar tumour) are conspicuous by their absence. The course of the disease, too, is different in the two cases; it is much more chronic in ataxic paraplegia than in the great majority of

cases of tumour of the cerebellum. The age often affords important information. Cerebellar tumours are much more common in children than in adults. The loss of sexual power and aching pains in the lower part of the back, which are present in a large proportion of the cases of ataxic paraplegia, are wanting in cases of cerebellar disease.

Prognosis.—The prognosis as regards arrest and cure is very unfavourable, as regards duration usually good. Most cases last for quite a number of years, the patient ultimately becoming unable to walk from the increased rigidity and weakness in the lower extremities.

Treatment.—The same measures which have been previously recommended for the treatment of the ordinary form of locomotor ataxia and of primary lateral sclerosis should be employed.

LECTURE XXIII

SYRINGOMYELIA

THE next condition, Gentlemen, to which I wish to direct your attention has only been worked out of recent years. It is termed syringomyelia. In its fully developed and typical form,¹ it is characterised *pathologically* by the presence in the spinal cord of cavities surrounded by gliomatous tissue; and *clinically* by (1) a peculiar derangement of sensation, viz., diminution or abolition of the sensibility to heat and cold and to pain, the ordinary tactile sensibility being retained; (2) in most cases, by localised muscular atrophy, which resembles more or less closely that characteristic of the Aran-Duchenne type; and (3) in many cases, by vasomotor and trophic disturbances, the exact nature of which I shall presently describe in more detail.

My object in directing attention to syringomyelia at this stage of the course is this, that it may be regarded as a focal lesion of the central grey matter.

In previous lectures, we have considered focal lesions of:—

1. *The anterior horn*,² the essential clinical features of which are:—(a) loss of motor power, (b) flaccidity and atrophy of the paralysed muscles, (c) abolition of the deep reflexes, and (d) the absence of any affection of sensation and of the bladder or rectum.

2. *Of the crossed pyramidal tract*, the essential characteristic features of which are:—(a) loss of motor power, (b) rigidity and tension of the paralysed muscles, (c) exaggeration of the deep reflexes, and (d) the absence of marked atrophy, of the reaction of degeneration and of any affection of sensation.

¹ There is probably no real distinction between gliomatous proliferation in the grey matter and adjacent parts of the cord and syringomyelia (i.e. gliomatous proliferation with the presence of cavities in the gliomatous tissue).

² Under this head should be included focal lesions involving the anterior root-fibres as they pass through the anterior columns.

3. *Of the posterior columns* (the postero-external columns and the ascending degeneration which is associated with it in the postero-internal columns or columns of Goll), the essential characteristic features of which are:—(a) abolition of the deep reflexes, (b) lightning pains and other derangements of sensation, (c) inco-ordination, and (d) the absence of muscular atrophy and of the reaction of degeneration.

4. Now, syringomyelia may be considered as a fourth focal lesion, *a lesion of the central grey matter* of the spinal cord, the essential feature of which is slowly and gradually developed diminution or abolition of the sensibility to thermal and painful impressions, with, in many cases (since the adjacent anterior and posterior horns of grey matter are very frequently involved), slowly and gradually developed muscular atrophy and vasomotor and trophic derangements in the skin, nails, hair, bones, joints, etc.

The four focal lesions which I have just enumerated (including the lesions of the anterior column which involve the anterior root-fibres) practically include all of the focal and system diseases of the spinal cord which are attended with definite and distinct localising symptoms. Further, we know that localised lesions, both systematic and indiscriminate, which involve localised segments or parts of segments, may produce very localised disturbances of function. Witness, for example, the paralyses of the bladder and rectum which result from lesions of the grey matter in the sacral segments (in which the centres for the bladder and rectum are situated), and the alterations in the pupil which result from some localised lesions in the lower cervical and upper dorsal segments (cilio-spinal region).

I do not say that localised lesions of parts of the transverse section other than those which have just been enumerated may not produce definite and distinct symptoms, or that other tracts or areas of the transverse section may not perhaps be involved by definite and distinct system lesions; but in the present state of our knowledge, we are unable to differentiate and recognise localised affections of other parts of the transverse section by the symptoms which they present during life. At the bedside, we know little or nothing of the effects which lesions produce in parts of the transverse section other than the four localised

areas which I have just enumerated (anterior cornua and adjacent part of the anterior columns, crossed pyramidal tracts, posterior columns, and the central grey matter and adjacent part of the posterior cornua).

Let us now turn our attention to syringomyelia.

Frequency.—The condition is probably more common than most authorities seem at present to allow. In quite a number of cases in which the lesions characteristic of syringomyelia were discovered post mortem the presence of the condition was unsuspected during life. In some of these cases, there were no definite and distinct symptoms—the lesion was actually latent. In others, the characteristic symptoms were missed or overlooked. As I shall presently have to tell you, this is very apt to occur; for unless the different forms of sensation are carefully tested, the diminished or abolished sensibility to thermal and painful impressions, in localised areas of the body, may very easily pass unobserved.

Etiology and Morbid Anatomy.—In the great majority of cases in which cavities are found in the spinal cord, they are undoubtedly the result of congenital malformations or of the breaking down (disintegration) of gliomatous tissue which may be regarded as a congenital formation. In rare cases, cavities in the spinal cord are the result of the ordinary forms of myelitis. The existence of this form of myelitis—the *myélite-cavitaire* of French authors—has been much debated; but there can, I think, be no doubt whatever that it does undoubtedly occur. I have met with at least two cases in which the evidence on this point was quite conclusive. In one, a cavity of considerable size in the lateral column resulted from a traumatic myelitis (see fig. 128). In another, a localised cavity in the anterior horn of grey matter was the result of a poliomyelitis anterior acuta (see fig. 129). But it must, I think, be admitted that cavities due to this cause (excavating myelitis) are extremely rare; and, so far as we at present know, they have no special pathological or clinical significance. They are not, so far as I am aware, attended with any characteristic symptoms, other than the symptoms which necessarily attend a severe destructive lesion in the part of the spinal cord in which the cavity happens to be situated. The congenital cavities are the only ones which, for our present

purpose, are of importance. They are the cavities which give rise to the definite and distinct group of symptoms which constitute the clinical picture of syringomyelia.

And here it is important to emphasise the fact that these congenital cavities are invariably, so far as we at present know, situated in the midst of, i.e. surrounded by, a mass of embryonic (neuroglial or gliomatous) tissue which in its structure closely

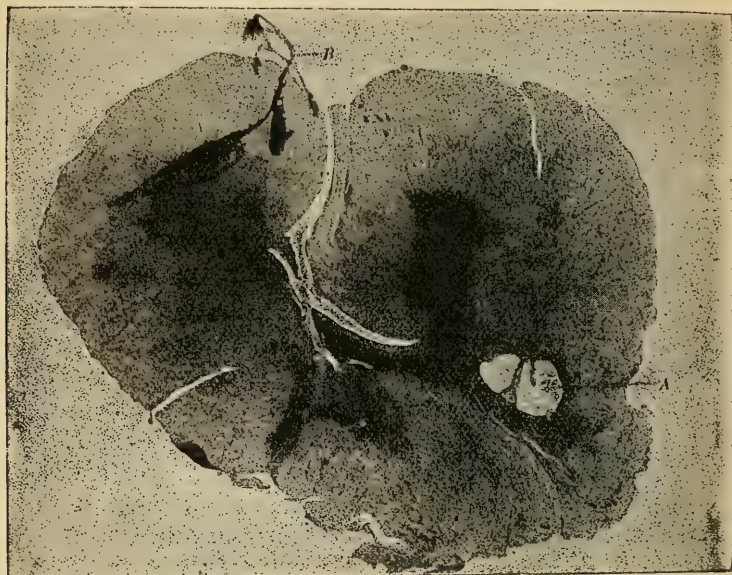


FIG. 128.—*Photograph of a transverse section through the cervical region of the spinal cord in a case of traumatic myelitis, showing a cavity in the lateral column.*

The letter *A* points to the cavity; the letter *B* to the membranes.

The cord was sent to me by Dr. Althaus; the case is referred to in his work on *Sclerosis of the Spinal Cord*, page 360.

corresponds to the cellular tissue of which the embryonic spinal cord is composed.

Quain describes three kinds of cells in the cord of the early embryo.¹ He says:—

‘The outermost layer of the embryonic cord, after the differentiation of the various kinds of cells above described has

taken place, is free from nuclei, and is composed of the partly reticulated, partly radially arranged external or attached extremities of the spongioblasts. This may be taken to represent the white matter of the cord at this stage (all the rest representing grey substance); but there are at first no nerve fibres in it, the only structures which can be at all compared to nerve fibres being the prolongations of the neuroblasts, and these lie either as arcuate fibres altogether in the outer part of the grey substance, or are passing out of the cord as the beginnings of the anterior roots from a mass of neuroblasts which forms the rudiment of the anterior cornu of grey matter. This mass constitutes in the human embryo of six weeks the chief portion of each half of the cord. It forms a considerable projection

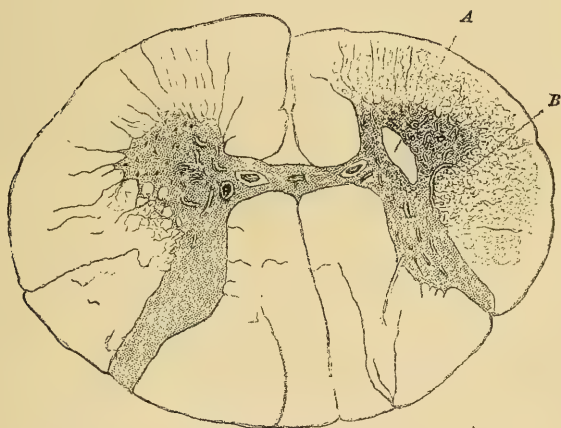


FIG. 129.—*Transverse section through the cervical region of the spinal cord in an old-standing case of poliomyelitis anterior acuta, showing a cavity (A) in the right anterior horn of grey matter. The adjacent part of the lateral column (B) is sclerosed; and the anterior and lateral columns are markedly wasted.*

which laterally almost reaches the surface, but ventrally is separated from it by a thickening of the external or radial zone, due to the appearance of longitudinally coursing nerve fibres within it; this is the beginning of the anterior white column. By this time, also, although to a rather less extent, the posterior white columns have, simultaneously with the posterior roots, begun to make their appearance on either side of the narrow dorsal part of the neural canal. There is, however, only a relatively thin layer of grey matter (neuroblasts) separating the

posterior white columns from the palisade-like lining of the canal, and as yet no sign of nerve fibres in the situation of the lateral columns, which are only represented by a thin layer of the radial myelospongium. The roof and floor of the canal are also quite thin and undeveloped.

‘At this period there is still no sign of either anterior or posterior (dorsal or ventral) fissures of the cord. These become formed as the cornua of the grey matter grow out from the central mass, and as the anterior and posterior white columns increase in extent. The anterior fissure is simply a cleft left between the enlarging lateral halves of the cords; the anterior commissure is formed across the bottom of the cleft, which is thereby separated from the central canal. As for the posterior fissure, it is uncertain whether it is in part formed from the dorsal portion of the constricted canal, which has become occupied by an ingrowth of pia mater, and converted into a mere septum of connective tissue, or whether this fissure with its connective tissue septum becomes formed independently of the central canal, which, as the fissure extends, gradually atrophies until it is eventually converted into the rudimentary epithelial tube which is persistent during life.’

Now, in the normal development of the spinal cord, the spinal canal which is so large in the foetus at the sixth week of its development becomes almost entirely obliterated. It remains merely as a narrow cavity, usually oval in form, situated in the centre of the posterior or grey commissure. But in some cases, owing to some defect of development, the exact causation of which we are at present altogether ignorant of, the obliteration is incomplete or partial. The result is that a cavity surrounded by a layer of embryonic tissue is present in the central parts of the spinal cord or the adjacent parts of the posterior horn of grey matter. In some cases, the cavity occupies a considerable portion of the posterior columns, and it not infrequently invades or compresses the anterior horn of grey matter. More than one of these cavities may be present in the same spinal cord. In many cases, the cavity communicates with the central canal at some point or another, and is in fact at places continuous with it; in others, the cavity seems entirely cut off from the central canal by a mass of gliomatous tissue in which they (the central canal and the pathological cavity) are both imbedded. To those cases in which the cavity seems to

consist merely of a greatly dilated central canal, the term hydromyelia has been applied. This condition is clearly congenital; when the dilatation of the central canal is great, the surrounding tissues of the cord may be atrophied, and, as it were, flattened out. In cases of this sort, there are often associated alterations in the brain, such for example as dilatation of the ventricles, absence of the cerebellum, etc. This condition of hydromyelia seems in some cases to be identical with the syringomyelia of the adult. When the dilatation of the central canal (the hydromyelia) is only slight in degree, it may be a condition of no importance; in cases of this kind, the dilatation is unattended with symptoms during life and is only discovered as an accidental condition after death.

In many of the cases of true syringomyelia in the adult, the cavities in the cord appear to be formed by the breaking down or disintegration of gliomatous tissue. In some cases, the lesion (the central glioma and the associated syringomyelia) extends throughout the whole length of the cord. (See fig. 130.) In others, and this appears to be much more common, the cavities are of limited extent from above downwards. The relative size of the cavities, and of the embryonic tissue which surrounds them, varies very considerably in different cases.

Further, you must remember that there is probably no real difference between those cases in which the gliomatous tissue in the centre of the spinal cord is in marked excess, in other words between cases of gliomatosis as the condition has been termed and cases in which cavities are present in the midst of the gliomatous tissue, in other words cases of syringo-myelia properly so termed.

As I shall presently point out, the clinical symptoms characteristic of syringomyelia are usually developed between the ages of twenty and thirty; but almost all observers are agreed that notwithstanding this fact, the lesion is usually a congenital one. It is universally believed that the gliomatous tissue which surrounds the cavities, and in some cases the cavities themselves, have existed from the time of birth.

And this leads me to say that the production of symptoms is the result of an increase either in the size of the cavities or of the gliomatous tissue which surrounds them. In other words, in those cases in which symptoms are developed, secondary

changes have taken place in the gliomatous tissue, the result of which is a proliferation and gradual increase of the glioma, the

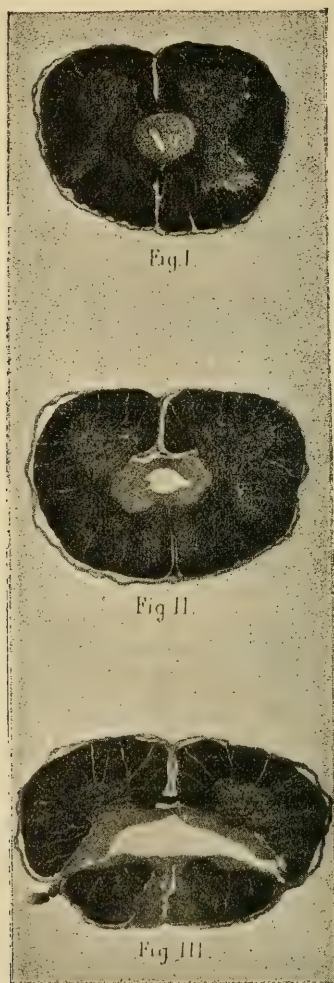


FIG. 130.—*Syringomyelia*.—(After Bruhl.)

Figs. 1 and 2 represent the condition in the lumbar and fig. 3 in the cervical enlargement.

enlargement of cavities which have previously been in existence, or the formation of new cavities, with the interruption of the

function of those parts of the spinal cord in which the glioma or the pathological cavity is situated or on which it encroaches and



FIG. 131.—*Transverse section through the cervical region of the spinal cord in a case of syringomyelia.*—(After Westphall.)

In the lower section, the cavity to which the letter *a* points is situated in the grey matter and adjacent portion of the posterior column.

presses. That this is the case is shown by the fact that the symptoms of syringomyelia are very rarely met with in children, and by the fact that cavities in the spinal cord, surrounded by a

gliomatous tissue have in several instances been met with in the spinal cords of children in whom the disease has been altogether latent, i.e. unattended by any symptoms during life. Cases in which the characteristic symptoms of syringomyelia are developed in childhood do, however, occasionally occur. An interesting example was recently brought before the Edinburgh Medico-Chirurgical Society by Dr. Bruce.

Syringomyelia is decidedly more common (probably twice as common) in males than in females.

I have said that in the great majority of cases, the cavities which are characteristic of syringomyelia are situated behind the cavity of the central canal; the lesion chiefly involves the grey matter and to a less extent the posterior columns. In some cases the central canal remains patent, and in fact in direct communication at some part or other, with the pathological cavity constituting the syringomyelia. In others, the central canal is obliterated, filled up with nuclear and cellular elements. But this, you will remember, is in no way peculiar. Obliteration of the central canal is of frequent occurrence. It is often seen in perfectly healthy cords and in cases in which there have been absolutely no symptoms of cord disease during life. The obliteration of the central canal seems in most cases to be entirely unattended with any injurious result. In some cases of this kind, the central canal is entirely obliterated throughout the whole cord; in others, the obliteration is only partial, and in these cases the unobliterated portions of the canal, which are situated immediately above or below the places at which it is choked up, may be somewhat larger than normal (dilated).

Further, it must be remembered that inflammatory changes in the grey matter closely adjoining the central canal, do not necessarily produce any injurious effect upon the canal itself. In the case which is represented in fig. 129 a severe inflammation of the anterior horn resulted in the formation of a cavity in the central part of the anterior horn. Now in that case the central canal in the same segment was absolutely healthy, and lined with a beautiful layer of ciliated epithelium. The shape of the central canal in this case is a point of interest. A small tailed projection passes from the round or oval portion downwards in the direction of the posterior median fissure. In some cases this tailed projection is of considerable size and indicates that the central canal

has been imperfectly closed by the normal obliterative process which occurs in the embryo.

Now, in some cases of syringomyelia this posterior, tailed portion is cut off from the anterior part and remains in the form of a cavity. This is one of the ways in which syringomyelia is produced.

The points, then, which I wish to emphasize are these:—(1) That the lesion which constitutes the pathological substratum of syringomyelia may exist for years without any symptoms; and (2) that in consequence of some irritation—it may be an inter-current illness or traumatic injury, for example—secondary pathological changes may be developed which lead to an outburst of pathological activity, in other words, to proliferation and increase of the gliomatous tissue, enlargement of the cavities or the formation of new cavities from disintegration of the gliomatous tissue with pressure upon, or destruction of, the adjacent cord tissues.

Further, hæmorrhagic extravasations may occur in the central glioma, and blood may be poured out into the syringomyelic cavities or into the adjacent tissues of the cord. When I come to speak of spinal hæmorrhage I shall have to tell you that blood is very rarely extravasated into the substance of the spinal cord in sufficient quantity to constitute a naked eye lesion, and that in all cases in which the hæmorrhage appears to be primary one should suspect the existence of a previous glioma. Exactly the same statement applies to intra-cerebral hæmorrhage in young persons—I exclude, of course, traumatic cases. Intra-cerebral hæmorrhage in young subjects and in cases in which there is neither arterial, cardiac, nor kidney disease, is not infrequently due to the rupture of the thin-walled vessels in a cerebral glioma. This fact should always be borne in mind in the investigation of cases of intra-cerebral hæmorrhage in early life, and in all cases in which the ordinary causes of intra-cerebral hæmorrhage (renal, cardiac and vascular lesions) are absent.

Remember, then, that the development of symptoms in cases of syringomyelia is usually delayed until adult life, and is then generally due to secondary pathological changes induced by the active growth or disintegration of the gliomatous tissue or to the enlargement of cavities which have existed in the spinal cord from the time of birth.

We see exactly the same sequence of pathological events in other organs and other tissues of the body. In the heart, for example, it is well known that a malformed valve (a pulmonary or aortic valve which has four or five cusps) is not infrequently the seat of acute endocarditis. Malformed valves seem to be more liable to be affected by inflammatory changes than normal valves. This no doubt is in many cases the result of mechanical causes—the malformation predisposes to mechanical injury and irritation. The significance of the occurrence of secondary endocarditis in a malformed aortic valve is less obvious than in the case of a malformed pulmonary valve, for the normal aortic valve is very frequently, as you all know, affected with endocarditis. But the normal pulmonary valve is very rarely indeed the seat of endocarditis; while, as I have just stated, a malformed pulmonary valve is not infrequently found to be affected with acute endocarditis.

This illustration will enable you better to realise and understand the important clinical fact that although the pathological condition which is the starting-point of syringomyelia not infrequently exists in a latent condition throughout childhood and youth, the clinical symptoms which characterise the condition and enable us to recognise or suspect the existence of the lesion are comparatively rarely developed until early adult life, usually between the ages of twenty and thirty, when active overgrowth or secondary pathological alterations (increased growth or disintegration of the neuroglial or embryonic tissue and enlargement of cavities) occur.

The cavities are most frequently situated in the lower cervical and upper dorsal regions of the spinal cord; hence the symptoms are usually developed in, or confined to, the upper extremities and the adjacent parts of the trunk.

In some cases, the lesion is unilateral; the symptoms are then confined to one limb (i.e. one arm) and the parts of the trunk adjacent to that limb. This was the case in the patient who is represented in Fig. 132, which, through the kindness of Dr. Alexis Thomson, I had the opportunity of examining and photographing.

In other cases, as I have already said, the lesion extends throughout a long tract of the spinal cord, indeed it may be throughout the whole length of the cord. In other cases again,

there may be several cavities separated from one another by unaffected portions of the cord, or, to speak more accurately, by masses of gliomatous tissue; for in these cases the embryonic tissue around the central canal is usually in excess throughout the whole length of the cord.

As I have already said, the cavity is usually situated in the central part of the cord, i.e. in the region of the central grey matter; and is either in direct continuation with, or just posterior to, the central canal. But remember that in a considerable proportion of cases the cavity involves the posterior column or the posterior cornu; that in other cases it encroaches upon the anterior cornu; and that in those cases in which the cavity is unusually large, it may press upon and interrupt the function of the nerve tracts in the lateral and anterior columns.

Inflammatory changes and hæmorrhagic extravasations in the gliomatous tissue are not uncommon. In some cases, secondary ascending or descending degenerations in the columns of Goll or in the crossed pyramidal tracts are developed. In those cases in which the anterior horn of grey matter is seriously involved, the anterior nerve roots which pass out from the affected area of motor grey matter and the motor nerve fibres which are continuous with them may be degenerated and atrophied.

In some cases of syringomyelia, an excess of gliomatous tissue has been present in the medulla oblongata about the floor of the 4th ventricle. In cases of this kind the nuclei of some of the cranial nerves may be involved.

Clinical History.—From the description of the pathological character and distribution of the lesion which I have just given you, you will easily see that the clinical symptoms which may be present in different cases of syringomyelia must necessarily be very variable. The onset is in most cases (probably in all cases) very slow and insidious.

The leading clinical symptom which is characteristic, though not by any means pathognomonic, of the condition, for it may occur in some other diseases (such, for example, as hysteria and leprosy), is a peculiar defect of the skin sensibility—a disassociation of the skin sensibility, as it has been termed. It is characterised by abolition or diminution of the sensibility of the skin

to heat and cold and to painful impressions; while the tactile sensibility of the skin and the muscular sense are, in the great majority of cases, unimpaired. The anæsthesia to thermal and painful impressions is usually, as I have already said, distributed over the upper limbs and adjacent parts of the trunk, neck and, it may be, the face. Though usually bilateral, it is sometimes unilateral. In some cases, the distribution of this characteristic sensory derangement is more extensive. It may affect the lower as well as the upper limbs, and the lower part of the trunk (abdomen, back). In a few cases, it has been one-sided in distribution.

The anæsthesia to thermal and painful impressions is not always co-equal in extent and degree. The anæsthesia to heat and cold is usually more marked and more extensive than the analgesia. In some cases, the thermal anæsthesia (to heat and cold) exists alone, the sensibility to painful impressions being, like the sensibility to tactile impressions, normal. In very rare cases, the patient can feel cold, but is unable to feel heat. In one case which has been reported the patient could perceive differences of temperature below 68° F.; above this point, thermal impressions were grossly perverted.

These defects of sensation to thermal and painful impressions are not usually distributed in the course of any particular nerve, but, like the sensory defects in some cases of anæsthetic leprosy and in hysteria, involve segments of the affected limb or limbs.

In consequence of the inability to perceive thermal and painful impressions, patients affected with syringomyelia sometimes burn themselves without feeling any pain.

In the earlier stages of the condition, the sensory defects may be very localised. There may, for example, merely be loss of thermal sensations over the hand. In cases of this sort, unless the condition of the whole skin to all forms of sensory impressions is very carefully investigated or unless the patient happens to burn the affected area without feeling pain, the presence of the defect is very likely to escape observation. This is one reason why cases of syringomyelia are so often overlooked in their early stages.

In the advanced stages of the disease, the tactile sensibility may, like the sensibility to thermal and painful impressions, be diminished or abolished; but this is rare. It only occurs in a

very limited number of cases, and is probably due to the extension of the lesion into the posterior-external column and posterior horn of grey matter, and to the involvement of the root-fibres as they enter the grey matter and the posterior horn.

In addition to the objective disturbance of sensation which I have just described, various subjective sensory symptoms may occur. Dull aching pains in the back and neck, and shooting pains in the upper extremities are sometimes the first symptoms which attract the attention of the patient and lead him to consult a physician. In some cases, the patient complains of feelings of heat or cold in the affected parts, of numbness, tingling, etc. In some cases, the conduction of thermal or painful impressions, instead of being abolished, is delayed.

LECTURE XXIV

SYRINGOMYELIA (*Continued*)

IN the last lecture, Gentlemen, we were considering the clinical history of syringomyelia. I had described the sensory alterations which constitute the most important and characteristic clinical features of the disease. Let me next direct your attention to the motor, vasomotor, and trophic alterations which are often present.

In a considerable proportion of cases of syringomyelia, probably in at least 50%, localised muscular atrophy is developed and is often the first condition to attract attention. In the great majority of cases, the atrophy commences in the small muscles of the thumb or hand, and corresponds more or less closely to the atrophy which is characteristic of the Aran-Duchenne type of progressive muscular atrophy. The muscular wasting is attended with fibrillary twitchings and the reaction of degeneration; the deep reflexes in the affected limb or limbs (the wrist and triceps jerks, for example, in the upper extremities) are often abolished.

As the lesion progresses, other muscles may become involved. In the case which is represented in Figs. 132, 135, and 136, the deltoid was completely atrophied, while the small muscles of the hand were unaffected. It is only in a comparatively small number of cases that the muscles of the lower extremities are involved. The muscles of the back probably suffer more frequently than is usually supposed.

In a comparatively small number of cases, spastic paraplegia is developed, doubtless as the result of secondary descending degeneration in the fibres of the crossed pyramidal tracts. It rarely happens that the lesion is sufficiently extensive to press upon and involve the pyramidal fibres. In those cases in which secondary descending degeneration is established in the

crossed pyramidal tracts, the spastic symptoms are usually developed in the latest stages of the disease. But this is not invariable. In one case described by Charcot, spastic paraplegia

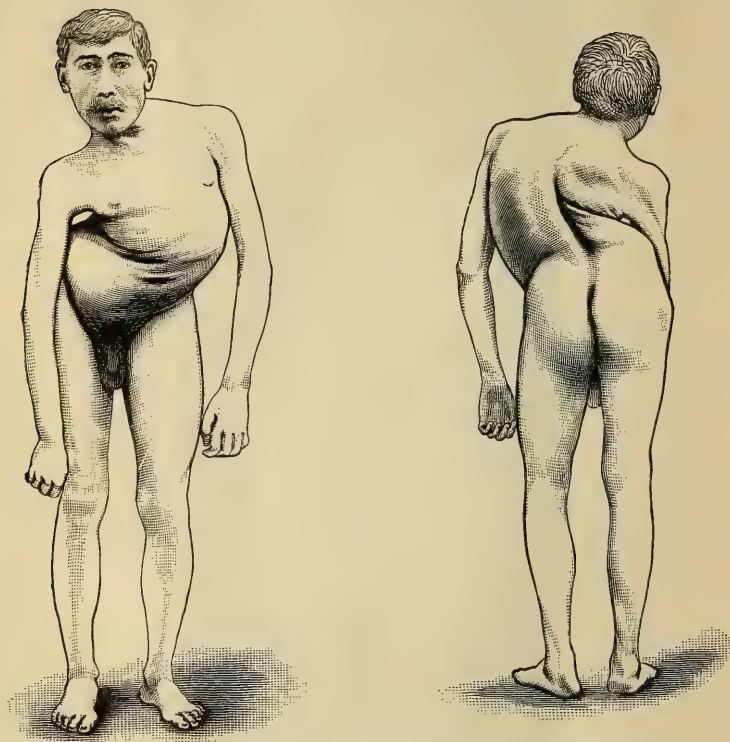


FIG. 132.—*Dr. Alexis Thomson's case of syringomyelia showing atrophy of the deltoid muscle.*

existed for several years before the characteristic symptoms indicative of syringomyelia were developed.

Deformities (curvatures) of the spinal column are often developed (see pp. 133 and 134).

In those cases in which the lesion involves the medulla oblongata and pons Varolii, muscles supplied by the motor



FIGS. 133 and 134.—*Extreme curvature of the spine in a case of syringomyelia.*
—(After D'Hallion).

nerves directly connected with these important parts of the central nervous system may become atrophied—the muscles supplied by the spinal accessory, hypoglossal, and facial nerves, for example. Atrophy of the tongue, difficulty in swallowing, paralysis of the larynx, are occasionally, though very rarely, developed in the later stages of the disease.

Vasomotor and trophic disturbances in the skin and its

appendages and in the bones and joints are comparatively common. In many cases, the hands are blue or red and cold; in some cases, swollen. The skin is in some cases thin and glossy, in others thick and horny. The nails may become brittle and cracked. Bullæ may form on the skin; ulcers which do not heal may be developed. Painless whitlows, which are so characteristic of Morvan's disease (a condition which, according to some observers, is identical with syringomyelia), are in some cases observed. The bones may become brittle; the joints may become diseased, and the joint lesions may present a very similar appearance to the joint affections of locomotor ataxia. In consequence of the elongation of the ligaments and the loosening of the capsule, the affected joint becomes more lax and mobile than normal; the head of the bone may become absorbed; in some cases villoid thickenings, or spiculæ of bone, are developed in the capsule. Like the joint affection of locomotor ataxia, these joint lesions are usually quite painless. They usually occur in the joints of the upper limb, especially the shoulder and elbow; in this respect they differ from the joint lesions of locomotor ataxia, which more frequently involve the joints of the lower extremity (the hip and knee). In the case which is represented in figs. 132, 135, and 136, the shoulder-joint was characteristically affected; the head of the bone was entirely absorbed, and the joint completely destroyed.

The peculiar character of the joint affection and its similarity to that which is characteristic of locomotor ataxia, is a point of great interest. It seems to suggest that the joint lesion of locomotor ataxia is probably due to implication of some part of the posterior grey matter, possibly of the nerve cells in the posterior horn.

The peculiar derangements of sensation are usually distributed over a much more extensive area than the muscular wasting or the trophic disturbances.

In some cases, the secretion of sweat over the affected areas of skin is abnormally profuse.

When the lesion involves the upper dorsal and lower cervical regions of the cord (cilio-spinal region), characteristic ocular changes may be developed. The palpebral fissure on the affected side may be narrowed, the eyeball retracted, and the pupil smaller than that on the opposite side. These appearances

were very marked in a patient who was under my care about a year ago. In that case, too, in which unilateral sweating was a conspicuous feature, the symptoms, though not perfectly typical, more closely resembled those of syringomyelia than any other



FIG. 135.—*Dr. Alexis Thomson's case of syringomyelia.*

The deltoid was completely atrophied and the head of the humerus absorbed. (See fig. 134.)

condition with which I am acquainted.

In some cases of syringomyelia, the patients are despondent, hypochondriacal, or melancholic.

The exact nature and distribution of the symptoms depend upon the position of the lesion in the spinal cord. As I have already pointed out, in the great majority of cases it is the

central grey matter which is first involved, the result being the peculiar sensory derangements, disassociated anaesthesia, as it has been termed—loss of sensibility to thermal and painful impressions, the tactile sensibility being unimpaired; and since in most

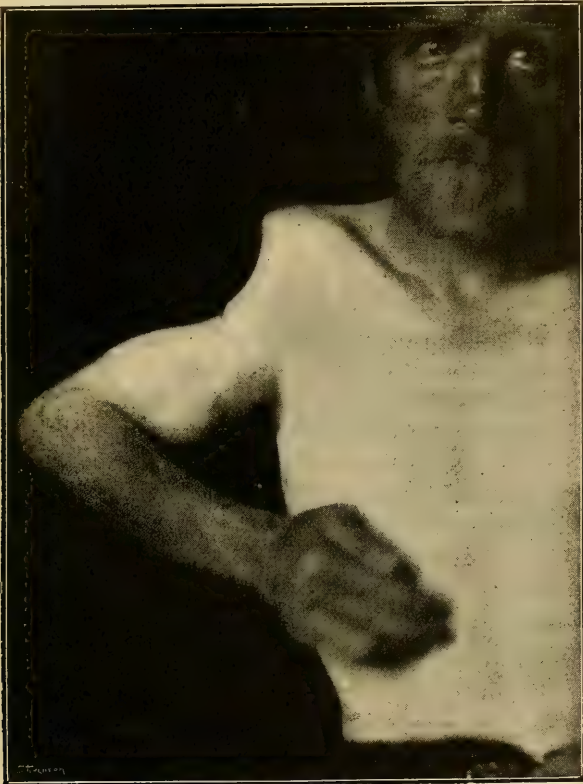


FIG. 136.—*Dr. Alexis Thomson's case of syringomyelia, showing the extreme mobility at the shoulder-joint.*

The head of the humerus has disappeared, the upper end of the shaft was blunt or truncated, and only attached by slack muscles and ligaments to the scapula. The case is recorded in the *Edinburgh Hospital Reports*, vol. ii. page 590.

cases the lower cervical and upper dorsal regions are first involved, the defect is usually first noticed in the upper limbs, or in one upper limb, if the lesion happens to be unilateral.

So long as the objective sensory derangements are the only

symptoms, the true nature of the case may easily escape observation. It is probable that in many cases of syringomyelia these symptoms had existed for some time—it may be for years—before the patient became aware that there was anything materially wrong with him. The occurrence of pains or other subjective sensory derangements, of localised muscular atrophy, or of vasomotor and trophic disturbances in the bones, joints, nails, etc., is usually a later development. As a rule, it is only after these symptoms are developed that the patient comes under the notice of the physician; but, as I have already pointed out, in some cases in which the disassociated anæsthesia is as yet the only symptom, the patient becomes conscious of the peculiar defect by burning the affected area of skin without feeling the burn.

After the secondary changes in the lesion and the production of symptoms have become conspicuous, the course of the disease is often, but by no means always, rapid. In a considerable number of the cases which have been reported, the patients have died within two or three years after the condition has been detected; but in many cases of syringomyelia the duration after the development of definite symptoms is probably much longer than this statement would imply.

Summary of the more important pathological and clinical features of syringomyelia.—To sum up:—Syringomyelia is a condition in which an excess of gliomatous tissue or cavities surrounded by gliomatous embryonic tissue are situated in the spinal cord. In the great majority of cases, the lesion involves the central grey matter and often implicates or encroaches upon the posterior columns, the posterior cornu, or the anterior horn of grey matter. In the majority of cases, the cavity or cavities are situated in the cervical and dorsal regions, usually the lower cervical and upper dorsal region of the cord; consequently, the symptoms are for the most part distributed over the upper limbs and adjacent parts of the neck and trunk.

In many cases, the lesion is latent (unattended by symptoms) until adult life, when, sometimes apparently as the result of traumatic injury, intercurrent illness or some local irritation, the gliomatous tissue surrounding the central canal actively proliferates and the cavities enlarge, or new cavities are formed in consequence of the disintegration and breaking down of the

gliomatous tissue. These secondary changes most frequently occur, and the symptoms are, therefore, most frequently developed, between the ages of twenty and thirty; but they may take place either at an earlier or a later period of life.

After the development of the symptoms, the course of the disease is usually progressive, but the rapidity of the progress and the duration of the case vary in different cases. The chief symptoms in the order of their frequency and diagnostic importance are:—

1. Diminution or loss of the sensibility of the skin to thermal and painful impressions (disassociated anæsthesia).

2. Subjective sensations of various kinds (pains in the back and affected limbs, sensations of heat, cold, etc.).

3. Localised muscular atrophy, usually affecting the small muscles of the hand (thenar and hypothenar), with fibrillary tremors and the reaction of degeneration.

4. Vasomotor derangements (blueness, redness, coldness, œdema).

5. Trophic derangements in the skin and its appendages, the bones and joints (glossy, thin, or thick skin; bullæ; increased secretion of sweat; painless whitlows; thickened or brittle condition of the nails; fragility of the bones, disorganisation of the shoulder, elbow, or other joints, etc.).

6. Deformity (scoliosis) of the spinal column.

7. Ocular alterations (contraction of the pupil, retraction of the eyeball, alterations in the size of the pupil).

Diagnosis.—In the earlier stages and in slight cases, the diagnosis is extremely difficult or impossible; for, as I have already pointed out, the characteristic sensory defects very readily pass unobserved. In many of the cases in which the lesions characteristic of syringomyelia have been found after death, the true nature of the case was unsuspected during life. The presence of the peculiar sensory defect characteristic of syringomyelia (loss of thermal and painful sensations, the tactile sensibility being unimpaired) should always raise the suspicion of syringomyelia. The possibility of the case being one of syringomyelia should also be kept in view when localised muscular atrophy is developed in the small muscles of the hand or in other muscles of the upper extremity (the deltoid, for

example), or when vasomotor or trophic lesions, and in particular the peculiar joint lesion which I have described, are present. In all cases of this description, the sensibility of the skin to all forms of stimuli (tactile, painful and thermal) should be very carefully and systematically tested.

Again, in the advanced stages of the diseases and in cases in which the lateral columns are pressed upon and involved—in other words, in those cases in which a condition of spastic paraplegia is present—the diagnosis is difficult, in consequence of the complicated character of the cord lesions. If, under such circumstances, the peculiar form of sensory impairment (disassociated anæsthesia) is found to be present, the presence of syringomyelia is almost certain, provided that a solid intramedullary tumour can be excluded.

There are several conditions—Morvan's disease, some cases of anæsthetic leprosy, Raynaud's disease, sclerodactylia—which more or less closely resemble syringomyelia. Indeed, some observers seem to think that all of these affections are mere varieties of the same condition. Such a conclusion seems to me, to say the least of it, premature. Although it must of course be allowed that in some cases of Morvan's disease and some cases which seemed to be closely allied to, if not identical with, anæsthetic leprosy, cavities have been found in the spinal cord.

The differential diagnosis of syringomyelia and of Morvan's disease.—In Morvan's disease, painless whitlows develop on the fingers. In that condition, the tactile sensibility is usually impaired, and in this respect it differs notably from syringomyelia; but whether Morvan's disease and syringomyelia are distinct conditions is still a disputed point. It seems certain that in many cases of Morvan's disease both the pathological and clinical features are identical with those characteristic of syringomyelia. In Morvan's disease, the same disassociation of sensory impressions has been observed, and cavities in the spinal cord have been detected after death; but in some of the cases of Morvan's disease which have been examined post mortem, the peripheral nerves have been more or less extensively inflamed. Further information is required in order to clear up the relationship which the two conditions bear to one another. It is possible that in some cases of Morvan's disease the condition is in reality syringomyelia, complicated by peripheral neuritis.

Raynaud's disease (symmetrical gangrene) and the rare affection of the skin which has been termed sclerodactylia, especially the latter, seem in some cases also to bear a close clinical relationship to syringomyelia.

The differential diagnosis of syringomyelia and the Aran-Duchenne type of progressive muscular atrophy.—This presents no difficulty, for although the muscular atrophy may affect the same muscles (small muscles of the hand and thumb or the deltoid), and may in the two conditions present identical features in respect to the presence of fibrillary tremors and the reaction of degeneration, the sensory condition is quite different.

The sensory disturbances and the vasomotor and trophic derangements, which are so characteristic of syringomyelia, do not occur in progressive muscular atrophy.

Further, in the Aran-Duchenne type of progressive muscular atrophy the muscular wasting is more frequently bilateral and symmetrical, and has a much greater tendency to spread and extend than the muscular wasting which is characteristic of syringomyelia.

The differential diagnosis of syringomyelia and anæsthetic leprosy.—As I have already pointed out, the clinical picture which the two diseases present is in some cases very similar,—in fact, almost identical. Indeed, some French writers have recently supposed that there is no real distinction between the two diseases; but this opinion is probably erroneous. In both conditions, there may be muscular atrophy localised or distributed in a similar way, very similar or identical derangements of sensation, and very similar trophic disturbances, such as ulceration of the fingers. But in leprosy, the tactile sensibility is, in the great majority of cases, impaired. In leprosy, the anæsthetic areas occur in patches, the margins of which are abruptly defined, and the areas of anæsthesia are usually more irregular in their distribution than in syringomyelia. In anæsthetic leprosy, the nerves are enlarged, pigmentation of the skin is often present, the inguinal glands are in some cases enlarged, and the patients have usually lived in a locality in which leprosy is endemic.

The differential diagnosis of syringomyelia and pachymeningitis cervicalis hypertrophica.—The two conditions bear some resemblance, inasmuch as muscular wasting and various sensory derangements in the upper extremities are characteristic of both.

But in pachymeningitis cervicalis hypertrophica, the course is more rapid, the bilateral character of the symptoms is more striking, the 'root-symptoms' are more prominent, and symptoms due to secondary degenerative changes in the crossed pyramidal tracts are usually much more marked. Again, in pachymeningitis cervicalis hypertrophica, shooting pains are much more prominent, and the tactile sensibility is markedly impaired—this is a most important point of difference. Further, in pachymeningitis cervicalis hypertrophica, the muscular atrophy is (usually) more uniformly distributed over the upper extremities; and the peculiar attitude of the hands which is represented in figs. 167 and 168, and spastic paraplegia with exaggeration of the deep reflexes are usually present.

The differential diagnosis of syringomyelia and intramedullary tumours.—A tumour which happens to be situated in, and limited to, the same parts of the transverse section of the cord which the lesion of syringomyelia usually occupies, would necessarily produce the same symptoms. Gliomatous tumours which take their origin from the gliomatous embryonic tissue around the central canal occupy this position. As I have already remarked, there is probably no real difference between this lesion (a central solid glioma) and syringomyelia (a glioma with cavities). My own opinion is that the two conditions are practically the same, and that the existence of cavities is in many cases largely accidental, although it must of course be admitted that in many cases the cavities (as well as the glioma) have existed from the time of birth, and are due to imperfect closure of the central canal.

Now, if solid central gliomatous tumours and syringomyelia are identical, it is obviously unnecessary to occupy time with the consideration of the differential diagnosis. But other forms of tumour are occasionally developed in the substance of the spinal cord.

Speaking generally, it may be stated that in the intra-medullary tumours the course is usually more rapid, and the development more continuous and progressive than in cases of syringomyelia. Again, in intramedullary tumours, the symptoms are seldom so sharply defined; they are usually more irregular in distribution and more widely diffused; the lateral columns are much more frequently invaded or pressed upon, and consequently paralysis and rigidity in the lower extremities or in one lower

extremity are much more frequently present, in other words, a tumour in the upper part of the cord, 'the seat of selection,' as I am in the habit of terming it, for syringomyelia, is much more likely to produce spastic paraplegia.

In those cases in which the tumour is situated in the lower dorsal or lumbar regions, the diagnosis is still more easily made, for the lesion which characterises syringomyelia is very rarely indeed confined to this part of the cord.

The differential diagnosis of syringomyelia and hysteria.—Charcot has pointed out that the sensory defects characteristic of syringomyelia and hysteria may be similar in this respect, that they are distributed over segments or divisions of the affected parts or limbs and are not confined to the area of distribution of any particular nerve. Further, he has shown that in some cases of hysteria the same form of sensory derangement which is so characteristic of syringomyelia (loss of the skin sensibility to thermal and painful impressions without any disturbance of the tactile sense) may be present; and that the same form of limitation of the visual fields which is characteristic of hysteria is occasionally observed in syringomyelia. Again, in rare cases of syringomyelia the sensory derangements are confined to, and distributed over, the whole or the greater part of one lateral half of the body—upper limb, side of the trunk, lower extremity, and it may be the side of the face and head on the same side. Again, in rare cases of hysteria, very definite vasomotor alterations are developed. I have at present under observation a young girl who has on several occasions manifested a most remarkable acute swelling, with lividity and temperature alterations, of the hands and forearms (first on one side, and then on the other). This vasomotor paralysis on each occasion appeared suddenly, and after lasting for several days or weeks as suddenly declined, under the application of the faradic current. The vasomotor paralysis was associated with loss of sensibility to tactile, painful and thermal impressions. The sensory paralysis was sharply limited to the affected forearm. There were absolutely no signs of organic disease, while there had been other symptoms (sudden loss of voice, etc.) suggestive of hysteria.

From these statements it is obvious that in some cases it must be extremely difficult to distinguish the two conditions. Fortunately, the difficulty in diagnosis does not often occur. In

the great majority of cases, there is no such close resemblance. It is only in quite exceptional cases that hysteria is likely to be confounded with syringomyelia, provided that the clinical phenomena are accurately and carefully observed.

Syringomyelia is more common in males, while hysteria is much more common in females. In syringomyelia, disassociated anæsthesia is the rule, while in the vast majority of cases of hysteria the tactile sensibility is affected as well as the sensibility to thermal and painful impressions. The presence of anæsthetic and hyperæsthetic areas is consequently a diagnostic point of great importance. The presence of associated symptoms of hysteria (fits, emotional disturbances, etc.) is another valuable point. In hysteria, localised muscular atrophy and the trophic derangements characteristic of syringomyelia very rarely if ever occur except as purely accidental complications, and such accidental complications are exceedingly rare.

In the few rare and doubtful cases, such as that which Charcot has reported, and to which I have just referred, in which the symptoms are insufficient to permit of a positive diagnosis, the true nature of the case can only perhaps be determined by observing the mode of development of the symptoms, the progress of the case, and the effects of treatment. In syringomyelia, the course is more progressive and the symptoms more fixed (less variable) than in hysteria. Fortunately, as I have already said, it is only in very rare cases of hysteria that the similarity to syringomyelia is so closely pictured. As a matter of practical fact the diagnosis can in most cases be made without difficulty.

The differential diagnosis of syringomyelia and peripheral neuritis.

—There is rarely any difficulty in deciding this question, unless indeed Morvan's disease, Raynaud's disease, anæsthetic leprosy and sclerodactylia are regarded as cases of peripheral neuritis. As I have already pointed out, the similarity between these conditions and syringomyelia is in some cases very close. But these conditions can hardly be regarded as cases of peripheral neuritis; for, even if it be allowed that the peripheral nerves are inflamed in these affections, their special and characteristic features distinguish them and separate them from the ordinary forms of peripheral neuritis.

Syringomyelia is distinguished from the ordinary forms of

peripheral neuritis by the peculiar character of the sensory disturbances (disassociated anæsthesia), and the fact that the sensory defects are not distributed in the areas of any particular nerve, nor bilaterally at the peripheral extremities of the nerves, i.e. over the hands and feet, as the sensory defects in cases of multiple peripheral neuritis are; and further, by the distribution and character of the motor, trophic and vasomotor derangements.

In the ordinary form of peripheral neuritis, the tactile sensibility is equally affected with, or more markedly involved than, the sensibility to painful and thermal impressions. In the ordinary form of peripheral neuritis, the sensory defects are limited to the areas of distribution of particular nerves or groups of nerves; while in the multiple form of peripheral neuritis, the sensory defects are bilateral and symmetrical, the fingers, hands, feet and toes being chiefly involved.

In the ordinary forms of peripheral neuritis, the motor and vasomotor defects are distributed and grouped in the same manner; and unless the lesion is very intense (and in such cases the other points to which I have already made reference are sufficient for the purpose of diagnosis), the vasomotor and trophic defects are seldom so well marked.

The differential diagnosis of syringomyelia and locomotor ataxia.—This presents no difficulty; for although the joint affection may be very similar or indeed identical in the two conditions, the associated symptoms are quite characteristic. A difficulty in the differential diagnosis of syringomyelia and locomotor ataxia is only likely to occur in those cases in which joint lesions are developed. In syringomyelia, the joint lesions are, with rare exceptions, only developed in the joints of the upper extremity (shoulder or elbow); whereas in locomotor ataxia the hip and knee are most frequently involved. In any case in which doubt existed, the nature of the associated symptoms would probably be sufficient to decide the diagnosis. In locomotor ataxia, the presence of lightning pains, loss of the knee-jerks, the Argyll Robertson condition of the pupil, Romberg's symptom, inco-ordination, urinary and sexual derangements, gastric or other crises, are the most important points. Further, in locomotor ataxia, localised muscular atrophy is rarely present, and the vasomotor and trophic disturbance are usually much less prominent than in syringomyelia. Again,

in locomotor ataxia the peculiar form of disassociated anæsthesia is rarely if ever present; while in those cases of syringomyelia in which painful phenomena are prominent, the pains are usually limited to the upper extremities and neck. The distribution of the sensory, motor, and vasomotor derangements is, in fact, a most important point. In locomotor ataxia, the characteristic symptoms (lightning pains, sensory derangements, inco-ordination, etc.) are usually limited to the lower parts of the body, or are much more marked in the lower than the upper extremities; whereas in syringomyelia the lower extremities are rarely involved except in the later stages of the case. Again, in locomotor ataxia the symptoms are bilaterally distributed; whereas in syringomyelia a unilateral distribution is by no means uncommon.

Prognosis.—This is very unfavourable. In the great majority of cases of syringomyelia in which distinct symptoms are present, the lesion continues to progress and to run steadily, but slowly, on to a fatal termination; in some cases, periods of remarkable temporary improvement have been observed. The course of the disease is, as we have seen, very variable. Each case must therefore be judged on its individual merits as regards the probable duration. The chief points which have to be taken into consideration in trying to form an opinion as to the probable duration of the case are:—(1) The extent of the lesion, as manifested by the nature, severity, extent, and distribution of the symptoms; and (2) the rate at which the symptoms appear to be developing.

Exhaustion, bedsores, cystitis, or some intercurrent pulmonary affection are in most cases the immediate cause of death.

Treatment.—So far as we know, little or nothing can be done to arrest the progress of the disease. Arsenic and iodide of potassium may be tried, but it is doubtful whether they exert any influence whatever upon the course of the lesion. In those cases in which the muscular atrophy is prominent, strychnine and electricity should be employed. The possibility of surgical interference—cutting down upon and tapping the cavity with a fine trocar—has been suggested, in those cases in which the locality of the lesion is well defined and the symptoms of increasing pressure on the spinal cord are well developed. It is

doubtful, however, whether this operation is likely to be attended with any real benefit. Further, it must be remembered that it is not unattended with risk. Sepsis may occur, and inflammatory changes in the gliomatous tissue which surrounds the cavity, or hæmorrhage may be developed.

Note on the cord changes in profound anæmia.—Lichtheim was the first to show that the nervous symptoms which are present in some cases of pernicious anæmia, are accompanied by definite histological changes in the spinal cord. The most frequent lesion appears to be a symmetrical degeneration (sclerosis) of the posterior columns, but in some cases other parts of the transverse section (crossed pyramidal tracts, etc.) are affected. Since I became acquainted with Lichtheim's observations, I have not had the opportunity of examining any case of pernicious anæmia post mortem; but for many years I have been well aware of the clinical fact that in that disease nervous symptoms suggestive of derangement of the functions both of the brain and spinal cord are sometimes present. In more than one of the cases which I published, now nearly twenty years ago, in illustration of the beneficial effects which I had obtained from the use of arsenic, the nervous (cord) symptoms were so prominent that they made a profound impression on my mind. Thus in reporting Case IV. (that of a man aged 43, who was admitted to the Royal Infirmary, Newcastle-on-Tyne, on March 18th, 1875, complaining of weakness and inability to walk and suffering from profound anæmia) I state (see *Edinburgh Medical Journal*, November 1887, page 418):—'He has great difficulty in walking, and his gait is very peculiar. He takes very long strides, and throws out his legs in an uncertain manner. The back is kept slightly arched, the head thrown back, the arms extended, one on either side of him, the palms being directed backwards, the forefinger and thumb of each hand approximated. When in bed he can move the legs in any direction. The reflex movements, on tickling the soles of the feet, are intense. The contractility of the muscles to the electric current is diminished. There is marked loss of grasping power in both hands. He is unable to approximate perfectly the forefinger and thumb, hence he cannot pick up a pencil from the table nor button his clothes. Sensibility to heat and cold is natural. He can perfectly localise impressions. He complains of pins and needles, and of numbness in both hands. The same sensations are felt in the legs, behind the knees.' And in commenting on the case, I state (page 419):—'The intensity of the reflex movements was remarkable; indeed, so great was it, that a condition of incomplete pseudo-paraplegia was produced, the debilitated condition of the patient partially contributing thereto. The intensity of reflex movement and the diminution of sensibility were no doubt due to the malnutrition of the nerve centres.' In another case (Case V.) there was also intense exaggeration of the reflexes.

The exact significance and causation of the cord lesions which Lichtheim and others have described in pernicious anæmia have not as yet been definitely elucidated. In most cases, they are probably the result of a widespread derangement of nutrition (directly due to the altered blood condition) in the nerve centres; and in this connection it is interesting to note, firstly, that in some cases of pernicious anæmia the cerebral symptoms (the result, I believe, of defective nutrition of the grey matter of the brain) are far more prominent than the spinal symptoms; and secondly, that with the removal and cure of the anæmia the nervous symptoms (both spinal and cerebral) disappear. In other cases, the cord changes are probably due to capillary hæmorrhages and resulting inflammatory and degenerative changes.

The subject was recently brought before the Royal Medical and Chirurgical Society by Dr. James Taylor. In the discussion which followed Dr. Taylor's communication, Dr. Gowers suggested that the cord lesions were probably due to some toxic substance in the blood. (See *Lancet*, March 30th, 1895, page 699.)

LECTURE XXV

ACUTE MYELITIS

Introductory Remarks.—The next subject, Gentlemen, which I propose to bring before your notice is myelitis. Inflammation of the spinal cord is a very important disease; for it is common, it not unfrequently proves fatal during the acute stage, and it very frequently leads to the production of permanent paraplegia; transverse myelitis is by far the most common cause of spastic paraplegia.

Classification of the various forms of myelitis.—Myelitis is a large subject. A number of different forms and varieties may be described. The acuteness of the inflammatory process, the extent and position of the lesion, and the cause of the inflammation may all be taken as the basis of classification.

The time classification; varieties of myelitis classified in accordance with the acuteness of the inflammatory process.—Myelitis may be either *acute*, *sub-acute*, or *chronic*. The distinction between these three forms is based upon the rapidity with which the inflammation is established and with which the symptoms are developed rather than upon the total duration of the disease and of the paralysis which it produces; for you must remember that acute myelitis, unlike many other acute inflammations (very unlike acute croupous pneumonia, for example), usually leaves some permanent damage, some paralysis, behind it. It is in many cases difficult or impossible to determine when the acute stage of acute myelitis is at an end; but there is seldom any real difficulty in deciding whether an inflammation of the spinal cord is acute or chronic, especially when a clear history is forthcoming, and when the physician has the opportunity of observing the case from its commencement.

In acute myelitis, the onset is rapid, and within a few hours

or days, a week or two at the very most, from the commencement of the attack, the greatest degree of development is reached.

In chronic myelitis, the onset is slow and gradual; and several, it may be many, weeks are required before the full intensity of the disease is reached.

Cases which occupy a mid-position between these two extremes are termed subacute.

It must of course be remembered that, in many of the cases in which the myelitis appears to be chronic and in which the symptoms continue to progress and develop from week to week, the morbid process is not merely a simple increase or development of the original lesion, but a series of fresh developments or inflammations, each of which (each new focus of inflammation) is an acute or subacute rather than a chronic myelitis.

Acute myelitis is a common disease; chronic myelitis is, comparatively speaking, a rare condition.

In the forms of myelitis which we are now about to consider, the lesion is essentially indiscriminate.

The clinical picture which different cases of myelitis present is very variable, for the symptoms depend, not merely upon the acuteness with which the morbid process is developed, but still more upon the extent of the inflammation, the height of the lesion in the cord, and the parts or areas of the transverse section which happen to be involved.

We have already considered one definite and distinct form of myelitis (poliomyelitis anterior acuta); and we have seen that it is not a mere variety of indiscriminate myelitis in which the lesion happens to be limited to the region of the anterior horn, but that it is a definite and distinct disease, probably due to a special form of irritant (perhaps a micro-organism or a ptomaine or toxine which results from a special micro-organism), which is prone to attack children.

Now, it is not improbable that some of the other forms of myelitis, which we at present classify in an arbitrary manner (basing our classification upon the extent and distribution of the inflammation and the acuteness with which it is developed), are also due to special irritants. If this is so, we may hope as our knowledge advances to classify the various forms of

myelitis on a scientific and etiological basis. Even at present, we attempt to make such a classification; for, as I shall afterwards point out, we speak of '*pressure*' myelitis, *syphilitic* myelitis, *traumatic* myelitis, and so on. But our knowledge of the exact causation of the myelitis is in many cases so imperfect that we have to be content with an arbitrary and non-scientific classification based either upon:—(1) Acuteness of the inflammatory process; and (2) the extent and distribution of the lesion in the cord and in the transverse section.

The anatomical classification; varieties of myelitis classified in accordance with the extent and position of the lesion.—Excluding poliomyelitis anterior acuta, which is a separate and distinct disease, the following varieties of myelitis may be distinguished, if we base our classification upon the extent and distribution of the lesion:—

1. *Transverse myelitis*. In this form, in which the inflammation involves the whole or the greater part of the transverse section of the cord, the vertical extent of the inflammation is, as a rule, small; the lesion does not extend, as the various forms of secondary ascending and descending degeneration do, in the form of a long, narrow strip through several segments of the cord, but as a short vertical band which implicates the whole or the greater part of the transverse section in one or two segments. The lumbar and lower dorsal segments are much more frequently affected than the cervical; I refer more especially to cases in which the myelitis is primary and intra-medullary, in which, in other words, the inflammation commences in the tissues of the cord. But you must remember that in many cases of transverse myelitis, the inflammation is secondary to an extra-medullary lesion (Pott's disease of the vertebræ, fracture or dislocation of the spine, a tumour on the surface of the cord, etc.). In those cases in which the myelitis is secondary to an extra-medullary lesion, the cervical region is not unfrequently affected (*i.e.* much more frequently affected than in the primary intra-medullary form).

A transverse myelitis in the dorsal or lumbar region produces the common form of paraplegia in which the lower extremities are affected. A transverse myelitis in the cervical region produces the rare form of paraplegia which we term cervical paraplegia; in it, all four limbs are paralysed.

The fact that acute transverse myelitis affects the lower dorsal and lumbar regions of the cord so much more frequently than the cervical region, is probably due to the comparative difficulty with which the lower end of the spinal cord is supplied with blood. I will refer to this point in more detail presently.

2. *Localised or focal myelitis.* In this form, the area of inflammation is limited to a part of the transverse section and the vertical extent of the lesion is small.

3. *Diffuse or general myelitis.* In this form, a large extent of the cord, both in the transverse and vertical directions, may be involved.

4. *Central myelitis.* In this form, the inflammation is chiefly limited to the central grey matter.

5. *Disseminated myelitis.* In this form, a number of separate foci of inflammation are scattered here and there throughout the cord or a portion of the cord; the vertical and transverse extent of each individual patch is small.

6. *Peripheral or annular myelitis.* In this form, which is probably always secondary to meningitis, the inflammation involves the superficial (surface) parts of the cord and may extend in the form of a ring round the whole thickness of the cord.

The etiological classification; varieties of myelitis classified in accordance with the cause of the inflammation.—The scientific or etiological classification of myelitis, which, as I have just told you, is as yet very imperfect, is of great importance for the purposes of prognosis and treatment.

In some cases, myelitis is due to traumatic injury (crushing of the cord by fractured or dislocated vertebræ, wounds, blows on the back, etc.); in other cases, to local vascular changes; in others, to the extension of an inflammatory process from adjacent structures (membranes, bones, etc.); and in others, to the presence of toxic substances in the blood. We may consequently divide cases of myelitis into *traumatic*; *idiopathic*; *rheumatic* (due to cold and wet); *syphilitic*; *diphtheritic*; *varioid*; *scarlatinal*; *compression myelitis*, and so on.

Now, speaking generally, we may say that in those cases in which the myelitis is limited and localised, as it is for example in transverse myelitis, the inflammation is usually due to vascular

changes (such as embolic or thrombotic plugging of the vessels), to traumatic injury, to compression or to the extension to the cord of an adjacent inflammation; whereas in those cases in which the myelitis is diffused and generalised, and in which it tends to extend throughout the whole cord, or in which several separate foci are produced in different parts of the cord simultaneously or in rapid succession, the inflammation is probably due to a toxic irritant carried to the cord by the blood. The irritant is in some cases a toxine or ptomaine derived from micro-organisms. This is certainly the case in diphtheritic myelitis, and in all probability in cases of myelitis following smallpox, scarlet fever, measles, influenza, and other diseases due to micro-organisms.

In others, the irritant is a chemical substance (such as alcohol or lead) introduced into the body from without. It seems certain that in some cases of alcoholic paralysis and lead palsy, inflammatory changes are present in the spinal cord as well as in the peripheral nerves.

From these statements you will see that myelitis is a very large subject, and that there are many different forms and varieties of the disease. I shall not attempt to describe each one of these individual forms in detail. The time at our disposal is quite insufficient for that purpose; and a systematic description of each of the varieties would only confuse you; the clinical differences which the different varieties present can only be satisfactorily and intelligently studied at the bedside when you have the living illustration before you. What I propose to do is to give you a general description of the pathological and clinical changes which are characteristic of myelitis—which show that the spinal cord is inflamed. I propose to take transverse myelitis as the type. By means of this general description, you will be able to differentiate myelitis from meningitis and the other conditions with which it is apt to be confounded. After you have obtained an intelligent knowledge of transverse myelitis, and therefore of myelitis generally, you will have no difficulty in understanding the clinical features which the other forms of myelitis (focal, disseminated, etc.) present. The detailed consideration which we have already given to poliomyelitis anterior acuta, primary sclerosis of the crossed pyramidal tracts, locomotor

ataxia and syringomyelia has made you familiar with the clinical symptoms and signs which result from lesions in the anterior cornua, crossed pyramidal tracts, posterior columns and central grey matter—the regions or areas of the cord which are of chief clinical importance. In myelitis, the lesion is indiscriminate; it invades at hap-hazard, as it were, the same areas and tracts which, in the diseases which we have so far considered, are invaded in a separate or systematic manner. Knowing the symptoms which result from lesions which are limited to the individual areas and tracts, you will readily appreciate and understand the symptoms which result from lesions (the different forms of myelitis) in which several of these areas or tracts are implicated, in which, in other words, the clinical picture is more complicated and composite.

Let us now consider acute myelitis in detail.

ACUTE MYELITIS

Morbid Anatomy.—The appearances which the spinal cord presents in different cases of myelitis are very variable; they depend upon the extent and severity of the inflammation, the length of time that the inflammation has lasted, and also to some extent upon the period at which the spinal cord is examined after death. (Post-mortem changes are rapidly produced in the delicate nerve tissues, and especially in nervous tissues which are softened or inflamed. In order to eliminate the possibility of post-mortem errors, it is advisable to examine cases of myelitis as soon after death as possible.)

The pathological changes which are met with in cases of indiscriminate myelitis are very similar to those which characterise poliomyelitis anterior acuta; and since you are familiar with these changes, I need not go into great details.

The cord may be abnormally soft at the seat of the inflammation. This is the only perceptible *external* change, unless of course the membranes are inflamed or the myelitis is due to an extra-medullary lesion (fracture or dislocation of the vertebræ, a tumour, etc.). When the cord is cut across, the appearances indicative of inflammation are more apparent.

Three stages of the inflammatory process are usually described.

In the *first* stage, that of *inflammatory engorgement, congestion and stasis*, the affected portion of cord usually appears to the naked eye abnormally red and hyperæmic; it swells up above the level of the adjacent healthy tissue; it is somewhat softer than natural. The congestion and hyperæmia are not always visible to the naked eye; in more than one case in which the cord was crushed by a fracture or dislocation of the vertebræ and in a condition of 'white softening,' I have found on microscopic examination all the characteristic appearances of acute inflammation.

In those cases in which the inflammatory changes are well marked, it may be impossible to distinguish by the naked eye the grey from the white matter. When the inflammation is less severe, the sharp outline between the grey and the white matter may be blurred and indistinct.

On microscopical examination, the vessels are found to be distended with blood; their lymphatic sheaths may be crowded with leucocytes; and capillary hæmorrhages may be scattered through the inflamed tissue. (See figs. 137 and 138.) The axis-cylinders and nerve cells are, even in this early stage of the inflammation, hypertrophied. (See figs. 139 and 140.) The swelling of the axis-cylinders is highly characteristic of myelitis. The connective tissue elements are commencing to proliferate, the stellate corpuscles (Deiters' cells) being unusually prominent. In the softened parts in which the nerve elements are breaking down, compound granule corpuscles and oil globules are seen.

In the *second* stage, that of *exudation and softening*, the redness and hyperæmia may have in great part disappeared, but the softening is usually much more marked; the affected parts of the cord may be completely disorganised and liquefied. Under the microscope, the nerve elements are seen to be broken down; compound granule cells and oil globules are abundant. When the softening is considerable, it may be impossible to harden and cut the affected portion of the cord in the ordinary manner. Under such circumstances, small portions of the softened tissue should be stained with aniline blue-black, picrocarmine, or osmic acid, mounted in glycerine and examined in the fresh state.



FIG. 137.—*Transverse section through the anterior horn of grey matter in a case of myelitis, showing engorgement of the blood-vessels.*

a, a, enlarged blood-vessels in the anterior horn; *b*, anterior median fissure; *c*, posterior median septum. I am indebted to Dr. Dickinson of Liverpool for the portion of cord from which these sections were made.



FIG. 138.—*Transverse section through the anterior horn of grey matter in a case of myelitis, showing great dilatation and engorgement of the blood-vessels and extravasation of leucocytes into the nervous tissue.*

a, a, dilated and engorged blood-vessels; *b*, extravasation of leucocytes.

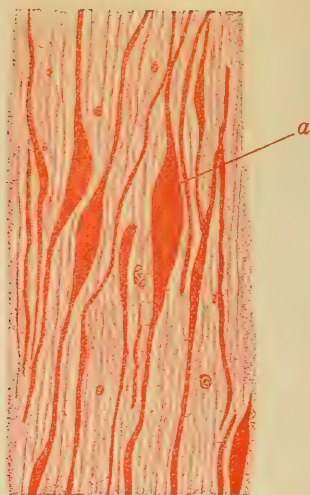


FIG. 139.—Longitudinal section through the spinal cord in myelitis, showing hypertrophy of the axis-cylinders.—(After Charcot.)

a, largest axis-cylinder.

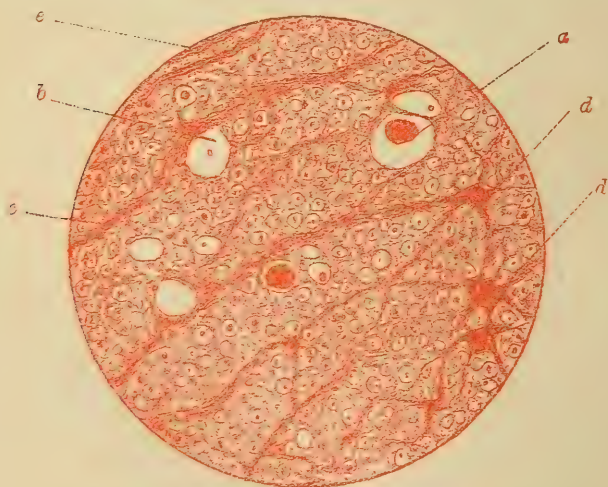


FIG. 140.—Transverse section through the dorsal region of the spinal cord in a case of chronic myelitis from compression. Stained with carmine, mounted in dammar, and magnified about 250 diameters.

a, hypertrophied axis-cylinder seen in transverse section; *b*, dilated nerve tube containing a normal axis-cylinder; *c*, sclerotic tissue containing numerous nuclei; *d, d*, Deiters' cells; *e*, blood-vessel with thickened walls.

Softening—inflammatory softening—is the essential naked-eye feature of acute myelitis; while enlargement (swelling) of the axis-cylinders is the most characteristic microscopic change.¹

In the *third* stage, that of *absorption and cicatrisation* the softened material is gradually removed. The affected tissue may be firmer than normal (sclerosed). Occasionally after the absorption of the inflammatory products and the process of cicatrisation are completed, a cyst remains.

On *microscopical examination*, the compound granular corpuscles and oil globules have now in great part, or entirely, disappeared; the connective tissue trabeculae are thickened (see fig. 140); the connective tissue cells are large and prominent; the blood vessels in the affected part are sometimes enormously dilated and their walls hypertrophied. The central canal is often filled with small round cells; but you must remember that this change is not peculiar to myelitis, indeed it has been present in the majority of adult cords (both healthy and diseased) which I have examined.

In some cases of myelitis, the nerve cells become vacuolated;

¹ It is often extremely difficult to distinguish simple (non-inflammatory) softening from the softening due to inflammation.

Simple (non-inflammatory) softening probably depends in the great majority of cases upon arrested or defective blood supply. Any alteration of the arterial walls, which narrows the vascular canal, will tend to produce it. In some cases, plugging (embolism or thrombosis) of the vessels going to the softened part can be demonstrated, but such cases constitute only a small proportion of the whole.

Erb states that simple (non-inflammatory) softening occasionally results from slow compression of the cord.

The microscopical examination of the softened parts is the only means by which we can distinguish simple softening from the inflammatory variety. Erb gives the following concise account of the microscopic appearances in the two conditions:—

‘Wherever we find a large number of cells containing fat granules, tensely distended blood-vessels, numerous young cells, increase of the interstitial tissue, swollen axis-cylinders, etc., we are justified in assuming positively the inflammatory character of the process. On the other hand, when these characteristics are wanting, and we only find simply swollen and disintegrated nerve fibres, ganglion-cells in a state of glassy swelling, a few cellular elements and fat-granule cells, and a small quantity of fatty detritus, we must diagnose a simple softening. Further and searching investigations of this point are, however, very much to be desired.’—Ziemssen’s *Cyclopaedia of Medicine*, vol. xiii. p. 470.

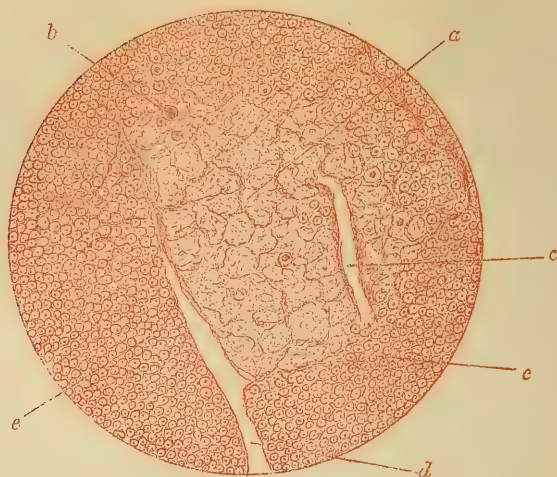


FIG. 141.—Transverse section through a portion of the posterior column, showing a patch of chronic myelitis. (\times about 150 diameters.)

a, patch of myelitis; *b*, enlarged axis-cylinder at the border of the patch; *c*, dilated blood-vessel with thickened walls; *d*, posterior median septum, which is a fissure in this case; *e*, *e*, healthy nerve tissue surrounding the lesion.



FIG. 142.—Transverse section through the cervical enlargement in a case of chronic myelitis, showing patches of miliary sclerosis (stained black with osmic acid), scattered all through the grey and white matter; the walls of the blood-vessels are also infiltrated here and there with the hyaloid material, of which the patches of miliary sclerosis are composed.

a, *a*, patches of miliary sclerosis in the grey, and *b*, *b*, in the white matter; *c*, *c*, blood-vessels, some of their walls are infiltrated and stained black with the osmic acid.

in others, the condition which has been termed miliary sclerosis is developed (see fig. 142).¹

To sum up. In slight cases of myelitis, the more important microscopical changes are:—Engorgement of the blood-vessels; extravasation of white and red blood corpuscles into the lymphatic sheaths of the engorged and dilated blood-vessels and into the inflamed nerve tissue; swelling and enlargement of the axis-cylinders and of the nerve cells; the production of compound granule corpuscles, and corpora amylacea; the proliferation of the connective elements and especially of the ‘spider-cells.’

In those cases in which the inflammation is severe, the tissues of the spinal cord may be extensively destroyed, softened or liquefied.

After the acute stage subsides, the inflammatory products are absorbed and sclerotic cicatricial changes are produced. Occasionally, when the myelitis is very severe and the destruction of tissue considerable, a small cystic cavity may ultimately be produced at the seat of the former softening.

¹ In hardened specimens, a glassy, colloid-like exudation is frequently met with; it surrounds the blood-vessels and infiltrates the tissues; the nerve cells of the anterior cornu may be greatly swollen and distended by this finely-granular, glistening material; small, round masses of the same material are sometimes seen in the interior of the blood-vessels. This glassy swelling of the nerve cells must be carefully distinguished from the inflammatory hypertrophy which Charcot has described. In the true inflammatory swelling, the enlarged nerve cells stain deeply with carmine; in the glassy swelling, the swollen cells are unaffected by that reagent, osmic acid stains the hyaline material a blue-black, methyl aniline a rose-pink colour; in true hypertrophy, the enlarged cells are unaffected by these reagents. The exact significance of this hyaline change is doubtful, but so far as I can form a judgment, it occurs in the great majority of cases of myelitis, both acute and chronic. Some good observers think that it results from the mode of preparation, and that it is due to the action of spirit. I have not been able to satisfy myself that it is entirely an artificial product. While I am prepared to allow that hardening in spirit favours its production, or as some observers would perhaps say, its demonstration, I have satisfied myself that the presence of miliary sclerosis is in most cases indicative of a diseased state of the tissue. Take, for example, two cords, one of which is perfectly healthy and the other affected with myelitis; harden them both in the same way, using some spirit as part of the hardening fluid; examine both cords at the end of six weeks, miliary sclerosis will, so far as I have been able to observe, be developed to a much greater degree in the diseased than in the healthy cord. Further, Dr. Tuke informs me that he has frequently met with patches of miliary sclerosis in brains which have been entirely hardened in Müller’s fluid; he is quite convinced that spirit is not necessary for the production of the lesion.

You must also remember that secondary ascending or descending degenerations are a frequent result of myelitis. In cases of transverse myelitis (provided of course that the patient survives a sufficient length of time for the production of the degenerative process), secondary ascending degeneration in the columns of Goll, above the lesion, and secondary descending degeneration in the crossed and direct pyramidal tracts, below the lesion, are conspicuous pathological features. The descending degeneration in the crossed pyramidal tracts has important clinical bearings, as we shall presently see.

If the myelitis should involve the region of the anterior horn, the root-fibres and the anterior roots of the peripheral nerves which are attached to the affected segment or segments of the cord present the degenerative changes which I have already described as such conspicuous features of poliomyelitis anterior acuta.

Further, I must ask you to observe that in some cases of acute myelitis, the inflammatory process involves the spinal membranes as well as the cord; meningo-myelitis is by no means uncommon.

Pathological Physiology.—Let us now direct our attention to the pathological physiology. In indiscriminate myelitis, as in poliomyelitis anterior acuta, the essential effect of the lesion will necessarily be to interrupt the function of the part of the cord which is involved in the inflammatory process. The rapidity with which the function of the inflamed tissue is arrested will, of course, depend upon the acuteness with which the inflammation is established; and the extent and severity of the symptoms (motor and sensory paralysis or impairment) will depend upon the acuteness and severity of the inflammatory process, and upon its extent and distribution.

Let us take as an example a case of acute transverse myelitis, and let us suppose that the inflammation involves one of the dorsal segments. The effect of the inflammation will be to rapidly or acutely interrupt or arrest the function of that segment.

Now, we have seen in connection with the physiology of the spinal cord that a spinal segment should be regarded:—

(1) As a *conducting medium* through which the great (*sensory* and *motor*) nerve tracts pass which connect the brain

above with the parts of the body which are situated below that segment.

A transverse myelitis which completely interrupts the functions of the segment (say one of the dorsal segments) will therefore produce complete motor and sensory paralysis in the parts below the segment (i.e. roughly speaking, and without going into too much detail, in the lower extremities).

And (2) *as a central organ*, connected with a localised part of the body to which the fibres of its anterior and posterior nerve roots are distributed or attached.

A transverse myelitis which completely interrupts the function of the segment (say in one of the dorsal segments) will therefore produce (a) motor and sensory paralysis in the parts of the body directly connected with that segment; (b) abolition of the reflexes which pass through the segment, and (c), if the inflammation is severe and if it involves the anterior cornu, rapid atrophy and the reaction of degeneration in the muscles with which the anterior root fibres of the segment are connected.

I should further say that the interruption of function and the destruction of the nervous tissue is, in some cases of myelitis, preceded by, or at its commencement attended with, symptoms indicative of irritation. I shall refer to these irritative symptoms (which are seldom prominent and always of brief duration) in more detail when I come to speak of the clinical history.

It is very important to remember:—

Firstly, that the loss of function (the motor and sensory paralysis) which is so characteristic of myelitis may, like the paralysis in cases of poliomyelitis anterior acuta, be due to two causes, viz., (a) temporary suspension of function, the result of shock or the pressure of inflammatory exudation; and (b) destruction of the nervous tissues; and *Secondly*, that the loss of function (paralysis and anæsthesia), which is due to the former cause (shock and pressure), is merely temporary, and may be expected to disappear with the absorption of the inflammatory products; but that the loss of function which is due to the latter cause (destruction of tissue) will remain. But even in the latter case, a considerable amount of improvement may occur in the course of time; for nerve fibres are more easily restored than nerve cells. In those cases in which the inflammation is very severe and the destruction great, and in which liquefaction and

extensive softening of the nerve elements are produced, the amount of repair and restoration of function will necessarily be small; but even in these cases, provided only that the cord tissues were previously healthy, some, and it may be considerable, improvement ultimately takes place. I shall return to this point when I come to the prognosis. Let me now only add that the sensory defects are more easily and more quickly repaired than the motor defects.

The essential points, then, in the pathological physiology of acute myelitis are:—

(1) That there is in some cases a preliminary stage in which irritative symptoms (seldom marked and always of brief duration) are combined with those of commencing destruction.

(2) That the essential result of an inflammation of the spinal cord is to produce a rapid interruption of function in the inflamed part; and that this may be either the result of (a) tissue destruction (in which case the resulting symptoms or some of them will remain), or (b) temporary arrest of function, due to shock and the pressure of exudation products (in which case the resulting symptoms will rapidly pass off). It is perhaps unnecessary to say that as a matter of fact both of these conditions are usually present; and that in some cases the one, while in some cases the other, is the chief pathological result.

(3) That the exact nature (extent and distribution) of the symptoms depends upon the extent and severity of the inflammation and the areas or parts of the cord which happen to be affected. In acute transverse myelitis, the functions of the inflamed segment, both as a centre and as a conducting medium, are interfered with and arrested, and in many cases more or less permanently abolished or destroyed.

(4) That in most cases (provided of course that the patient recovers from the immediate effects of the inflammation) there is considerable improvement; and that this may be due either to the absorption and removal of exudation products or to restoration of the deranged nerve elements (nerve tubes). Remember further, that the sensory nerve tubes, or at all events the sensory functions, are, speaking generally, more easily restored than the motor.

(5) That as a result, more especially of transverse myelitis, secondary descending degeneration in the crossed pyramidal

tracts is produced and it is attended with exactly the same results (symptoms) which we have already studied in connection with primary spastic paraplegia.

Etiology.—Let us now turn to the etiology. In speaking of the classification, I have told you, perhaps at too great length, that myelitis may be due to a number of different causes.

For the purposes of prognosis and treatment it is most important to remember, that in some cases the myelitis is primary or intra-medullary, and that in others it is a secondary result of some other (extra-medullary) lesion. The extra-medullary lesions which most frequently produce myelitis are Pott's disease of the vertebræ, fractures or dislocations of the spine, and the pressure of new growths or of inflammatory products upon the cord.

If we exclude the secondary forms, we find that (primary) myelitis is chiefly a disease of young and middle-aged adults. It is rare in children and in old people. I refer of course to the ordinary indiscriminate form, not to poliomyelitis anterior acuta. It is much more common in men than in women.

Anything which debilitates the body as a whole and the cord in particular may probably act as a predisposing cause of myelitis. Sexual excess and violent muscular efforts which throw a strain upon the tissues of the cord, appear in some cases to predispose to, or perhaps to actually produce, myelitis. In one case, which came under my notice a few years ago, in which a patient was making a good recovery from a transverse myelitis, a sudden relapse (and permanent paraplegia) seemed to be the direct result of a long walk. Now, if excessive muscular effort may re-excite inflammation in a spinal cord which is recovering from a myelitis, I see no reason why it may not, in some cases at all events, act as a predisposing or perhaps even as an exciting cause of myelitis in a cord which was not previously inflamed.

Exposure to cold and wet is one of the most frequent causes of acute myelitis. Several cases have come under my own notice in which an acute inflammation of the spinal cord was clearly due to this cause. In one case, a healthy man fell asleep on the damp grass; the following day a very acute myelitis developed. In another case, a healthy man lay out while in a

state of drunkenness on the wet ground; the next day he became paraplegic as the result of an acute transverse myelitis.

Direct injury to the back which does not produce any injury to the bone (any fracture or dislocation of the vertebræ) is occasionally though very rarely followed by acute myelitis. In some cases of this kind, the spinal symptoms are developed immediately or soon after the accident; in others, after an interval. I shall consider these cases in more detail when I come to speak of concussion of the spinal cord and the lesions and diseases of the spinal cord which result from traumatic injury.

Syphilis is, I believe, a not uncommon cause of myelitis; but there is much difference of opinion on this point. Several cases have come under my own observation in which both transverse and disseminated myelitis seemed to be due to syphilis. Acute myelitis is sometimes developed after measles, small-pox, typhoid fever, influenza, gonorrhœa, pyæmia and other 'germ diseases.' In cases of this kind the myelitis is usually disseminated or diffused. In the pyæmic cases, meningo-myelitis is probably more common than simple myelitis; in the gonorrhœal cases, which are very rare, and in some of the pyæmic cases, the septic irritant is perhaps carried to the spinal cord by the blood-vessels or lymphatics, or the inflammation travels (ascends) along the nerves which are attached to the sacral and lumbar segments of the cord.

Myelitis is sometimes apparently due to sudden arrest of the menstrual flow. In a case which I have recently had under my care in hospital, a girl aged 26 bathed in the sea while menstruating; the flow was immediately arrested; she felt giddy; gradually experienced difficulty in walking, and three months afterwards was completely paraplegic.

Clinical History. We have seen that the severity, extent and distribution of the inflammation are very different in different cases; these differences are reflected in the clinical picture which different cases of myelitis present. Let us take acute transverse myelitis—the most frequent form of myelitis—as our type. I have already told you that acute transverse myelitis is by far the most common cause of spastic paraplegia.

The outstanding symptoms which characterise acute trans-

verse myelitis are:—Motor paralysis of the paraplegic type; loss of sensation in the paralysed parts; paralysis of the bladder and rectum; and a tendency to the formation of bed-sores. In acute transverse myelitis, these symptoms develop rapidly, and the onset of the inflammation is usually attended with more or less fever and other symptoms of constitutional disturbance.

In the great majority of cases, the inflammation involves the lower part of the cord (the lower dorsal and lumbar segments). The cervical and upper dorsal segments are, comparatively speaking, very rarely affected. This is a point of great importance. Consequently, in the great majority of cases of transverse myelitis, the motor, sensory and other symptoms affect the lower and not the upper extremities. The dorsal segments are more frequently involved than the lumbar enlargement. The exact nature of the symptoms (the condition of the paralysed muscles, of the reflexes and the character of the bladder and rectal paralysis) depends upon the position of the lesion, the essential point being whether the lumbar enlargement—the part of the cord in which the trophic and reflex centres for the lower limbs and for the bladder and rectum are situated—is directly implicated or not. But I must leave the more detailed consideration of this point until the lecture to-morrow.

LECTURE XXVI

ACUTE MYELITIS (*Continued*)

IN the last lecture, Gentlemen, I had commenced to describe the clinical history of acute myelitis. We saw that the symptoms vary very considerably in different cases and depend upon the acuteness and severity of the inflammation and upon the extent and position of the lesion. In the most common form, transverse myelitis, which I propose to take as a type, the inflammation usually involves the dorsal or lumbar portion of the cord. The paralysis is consequently paraplegic in distribution; but the condition of the paralysed muscles, of the deep reflexes, and of the bladder and rectum present important differences, the essential point being whether the myelitis involves the trophic and reflex centres in the lumbar and sacral regions of the cord or not. But before describing these differences in detail, it may perhaps be well to refer more particularly to the mode of onset and the way in which the symptoms are developed.

Constitutional symptoms.—The nervous symptoms which result from derangement of the functions of the spinal cord may be preceded by, or developed simultaneously with, fever, quick pulse, malaise, restlessness and thirst—in short, the ordinary constitutional symptoms which attend any acute febrile process. In some cases, there is headache; in young patients (but in children ordinary indiscriminate myelitis is, you will remember, extremely rare), and in persons of high nerve susceptibility, so to speak, more marked cerebral symptoms, such as nocturnal delirium, muscular twitchings, or even an actual epileptic fit, may occur. But such cerebral disturbances are rare at the commencement of acute indiscriminate myelitis, although as we have seen they are by no means so uncommon in children at the commencement of poliomyelitis anterior acuta.

The severity of the initial fever and constitutional disturbance is very variable, but is more proportionate to the extent and severity of the spinal inflammation than in poliomyelitis anterior acuta. In that disease, the severity of the constitutional disturbance may, as we have seen, be quite disproportionate to the severity of the local mischief.

The degree of *febrile disturbance* varies greatly in different cases. In some, the temperature rises rapidly and attains a considerable height, 103° or 104° Fahr.; while the pulse may reach 120, 130, or even 160 in the minute. Headache, loss of appetite, thirst, and a feeling of malaise are, in cases of this kind, well marked. In other cases—and they constitute the majority—the elevation of temperature is slight and the general constitutional symptoms inconspicuous. The fever may continue for some time, but as a rule it quickly subsides. When acute bed-sores, acute cystitis, or pyelonephritis are developed, a fluctuating temperature and hectic or pyæmic symptoms may be present. This secondary fever must be carefully distinguished from the primary fever, which in many cases is slight or altogether absent.

In the more severe forms of acute myelitis, the onset may be attended with a well-marked rigor.

In other cases (and they probably constitute a considerable majority), the spinal symptoms are developed at the commencement of the case; in some cases, before the fever and constitutional disturbance are apparent.

It occasionally happens that the motor paralysis is so rapidly developed as to suggest the occurrence of spinal hæmorrhage; indeed it is probable that in some of these cases there is hæmorrhage, but the bleeding is the result and not the cause of the inflammation. I shall refer to this point again presently.

Irritative symptoms.—In some cases, the paralytic symptoms indicative of interruption of function and destruction of tissue are preceded by a brief stage of irritation; or perhaps it would be more correct to say that irritative phenomena are, in some cases, developed simultaneously with the early paralytic symptoms (numbness, loss of motor power in the legs, inability to empty the bladder, retention of urine, etc.). But the occurrence of these irritative phenomena is by no means constant; and even in those cases in which they do occur, they are rarely prominent,

unless, indeed, the membranes are also involved. Meningomyelitis is, as we shall afterwards see, comparatively common ; but in these cases, the inflammation of the cord is usually secondary to the inflammation of the membranes.

The irritative symptoms may be either sensory or motor. The symptoms indicative of sensory irritation are usually the most prominent. They consist of painful sensations of some kind or another, such as :—myalgic pains ; hyperæsthesia ; neuralgic pains shooting round the trunk or down the limbs in the areas of distribution of nerves attached to the affected segments of the cord ; girdle sensations, a feeling of constriction round the waist, etc. These symptoms indicative of sensory irritation are of considerable diagnostic importance, for they are rarely met in the functional derangements which are likely to be confounded with acute myelitis and acute meningitis.

Pain in the back is not a marked symptom in myelitis ; in fact, it is very usually absent and it is probably never severe, unless the myelitis is complicated by, or associated with, meningitis.

These irritative sensory phenomena are, as I have just remarked, usually associated with symptoms indicative of sensory impairment—numbness, pins and needles, more or less anæsthesia and analgesia. In the course of a few hours, the symptoms indicative of sensory irritation usually subside and the symptoms indicative of sensory paralysis become more marked.

In addition to the symptoms indicative of irritation of the sensory nerve apparatus, muscular twitchings, tremors, cramps and spasms, indicative of irritation of the motor nerve apparatus, may also be developed. They are seldom prominent in myelitis. As a rule, they are chiefly or only marked in the muscles which are connected with that portion of the cord which corresponds to the upper level of the lesion—with that part of the cord at which the inflammatory area abuts, so to speak, upon the healthy cord tissues. In short, the cramps, spasms, etc., are in most cases due to the irritation of motor fibres or cells which are not gravely implicated by the inflammatory process.

In most cases, the symptoms indicative of motor irritation are from the first associated with some symptoms indicative of motor paralysis (some loss of power in the legs and some

difficulty in urination). In the course of a few hours, the muscular twitchings and spasms usually subside and the motor paralysis becomes more marked.

Paralytic symptoms.—Now, the intensity of the paralytic symptoms depends, as I have so repeatedly told you, upon the severity of the inflammation and the exact area and extent of the transverse section of the cord which happens to be involved.

In most cases, I am speaking you remember of acute transverse myelitis, the motor and sensory paralysis below the lesion is complete, or at all events very marked, once the paralytic symptoms are fully developed. In those cases in which the inflammation is less severe and the whole transverse section of the cord is not invaded, the paralysis may, of course, be less extensive.

The motor paralysis is usually more marked than the sensory paralysis. In slight cases in which the motor paralysis is incomplete, the loss of sensation is often inconsiderable. Further, as we shall afterwards see, the sensory defects are, as a rule, more quickly recovered from than the motor defects.

I must here ask you to note that in acute transverse myelitis, as in poliomyelitis anterior acuta, the initial paralysis is not necessarily co-equal with the permanent paralysis. In the less severe cases, a considerable amount of the initial paralysis may pass off with the subsidence of shock and the absorption of the inflammatory products.

In severe cases, then, of acute transverse myelitis, involving the dorsal or lumbar region of the cord, motor paralysis (paraplegia) and anæsthesia in the lower extremities and in the lower parts of the trunk, are established, in the course of a few hours, or at most a day or two. Further, the bladder and rectum are usually paralysed, and in many cases bed-sores are rapidly developed.

The early and marked paralysis of the bladder and rectum and the development of bed-sores are diagnostic points of the greatest importance. They constitute some of the chief points of distinction between myelitis and meningitis, and between the indiscriminate form of myelitis which we are at present considering and poliomyelitis anterior acuta.

Now, if the patient be examined during this, the early stage of the disease, when the inflammation has been in existence for

some hours or days, and when the loss of motion and sensation and the paralysis of the bladder and rectum are well marked, it will generally be found that some symptoms of constitutional disturbance (fever, quick pulse, headache, and so on) are present. As a rule, the sensorium remains quite clear.

The condition of the paralysed muscles and the exact character of the paralysis of the bladder and rectum vary in different cases and depend upon the position of the lesion, the essential point being, as I have already insisted upon, whether the trophic and reflex centres connected with the lower limbs and with the bladder and rectum are directly implicated or not.

Let us take two illustrative cases, two cases of severe acute transverse myelitis, and let us suppose that, in one, the inflammation involves one or more of the segments of the cord in the mid or lower dorsal region; and, in the other, the segments composing the lumbar and sacral regions of the cord.

In both cases, there may be complete loss of motor power and of sensation in the lower extremities, paralysis of the bladder and rectum, and the formation of bed-sores.

But there are important differences. In the first place, the *extent and distribution of the motor paralysis and of the anæsthesia* is not exactly the same. In the former case the paralysis extends higher up the trunk and involves the abdominal and thoracic muscles; whereas in the latter case the abdominal muscles (except the lower parts of the rectus and internal oblique muscles which are supplied by the first lumbar nerve) and the thoracic muscles escape. The exact distribution of the paralysis in the abdominal and thoracic wall is in many cases determined with difficulty; but the upper limits of the anæsthesia and the condition of the abdominal reflex at different levels are readily ascertained.

In the second place, *the condition of the paralysed muscles* is very different. A transverse myelitis in the dorsal region does not implicate the reflex and trophic centres for the muscles of the lower extremities; consequently the paralysed muscles of the lower limbs do not undergo the rapid form of atrophy, though they may and usually do present some wasting (atrophy of disuse); they do not present the reaction of degeneration; and their reflexes are (after the initial effects of shock and inflammation have passed off) not abolished; on the contrary, they

become (in the course of a little time) exaggerated.¹ In short, a transverse myelitis in the dorsal region produces a condition of typical spastic paraplegia.

In the course of time the atrophy of disuse may in great part disappear, partly as the result of recovery and return of voluntary power and partly as the result of reflex stimulation. We have seen that in spastic paraplegia the muscles are being constantly exercised and put into contraction by reflex irritations passing from the periphery; and as I have repeatedly told you, a transverse myelitis in the dorsal region ultimately produces a condition of spastic paraplegia.

But a transverse myelitis of the lumbar enlargement, which destroys the trophic centres (the multipolar nerve cells) for the muscles of the lower extremities, produces not merely a paraplegia, but a paralysis in which there is rapid atrophy, the reaction of degeneration and abolition of the reflexes.

It is perhaps unnecessary to say that in those cases in which the lesion is partly dorsal and partly lumbar, or in which a portion only of the lumbar enlargement is involved, a combination of these two forms of paraplegia may be produced. In these mixed cases, which are more common than the cases in which the whole of the lumbar grey matter is involved, some of the paralysed muscles may undergo the rapid form of atrophy and present the reaction of degeneration, and some of the reflexes may be abolished while others may be exaggerated.

It must also be remembered that, if Dr. Charlton Bastian's view as to the effect which a complete transverse lesion of the cord has upon the condition of the reflexes is correct (see page 127), total and complete destruction of the whole thickness of the cord in the dorsal region will be attended with permanent abolition of the reflexes in the lower extremities. I have already considered this question in connection with spastic paraplegia. I need only now say that absolute and complete destruction of the whole thickness of the cord is very rarely if ever the result of an ordinary indiscriminate transverse myelitis; and as a matter of fact, a transverse myelitis in the dorsal region is almost invariably attended (after the initial effects of the shock and inflammation are recovered from) with marked exaggeration

¹ Exceptions to this statement are met with in some cases in which the transverse lesion is complete. (See page 127.)

of the deep reflexes (great exaggeration of the knee-jerks and ankle-clonus).

In the third place, *the paralysis of the bladder and rectum* may present important differences in the two cases. A severe transverse myelitis either in the dorsal or the lumbo-sacral regions of the cord, is attended with paralysis of the bladder and rectum. But a myelitis in the dorsal region does not directly implicate the reflex centres for the bladder and rectum. Consequently, after the primary effects of the shock and inflammation are recovered from, the sphincters are not completely paralysed. In the course of time, although the patient may (in consequence of the block to the upward passage of sensory impressions) receive no information as to the condition of the bladder, and (in consequence of the block to the downward passage of voluntary motor impulses) may be unable to exert any voluntary control over the condition of the bladder, either in the direction of accelerating and strengthening or of retarding the act of micturition, the bladder may still be emptied reflexly. There may be no dribbling and no retention. Urine may collect in the bladder and at periodic intervals (when the bladder becomes full) be discharged unconsciously. This condition of '*unconscious reflex micturition*,' as I term it, is only seen in some cases. In the early stages of a transverse myelitis, there may be dribbling from paralysis of the sphincter or retention from paralysis of the detrusor (expulsor) muscle. In many cases, after the initial effects of the lesion are recovered from, the detrusor remains paralysed or enfeebled.

When the reflex centre for the bladder, which is situated in the sacral region of the cord, is directly implicated and destroyed by the inflammatory process, the sphincter is permanently paralysed, and the urine constantly dribbles away. In cases of this kind, the patient loses all control over the bladder, and if the lower extremities are completely anæsthetic, he does not know that the water is dribbling away. This is a much more serious form of incontinence.

Between these two extremes, there are all degrees of difference. A transverse myelitis in the upper lumbar segments does not directly involve and destroy the reflex centres for the bladder and rectum.

In order that these urinary derangements may be clearly

understood it may perhaps be well to describe the nervous arrangements by which the expulsion and retention of urine are effected in more detail.

The vesical reflex.—Micturition is a complex process; the nervous and muscular arrangements concerned are as follows:—

The neck of the bladder is surrounded by circular muscular fibres—the sphincter vesicæ muscle—which are supposed to be maintained in a constant state of contraction. By virtue of the action of the sphincter, and, to a less degree, of the resistance of the elastic fibres of the urethra, the exit of urine is prevented. The contraction of the sphincter is due to the action of a tonic centre which is situated in those segments of the spinal cord which correspond to the 3d, 4th, and 5th sacral nerves. The action of the tonic centre can be inhibited, *i.e.* the sphincter can be relaxed, by a voluntary effort. The course of the inhibitory fibres in the cord is undetermined, but they probably pass through the lateral columns. Dr. Gowers supposes that the centre for the sphincter can be inhibited, *i.e.* the muscle can be relaxed, *reflexly*, by sensory stimuli from the mucous membrane of the bladder itself. Such a supposition would undoubtedly well explain some points of the process, and is adopted in the following description.

The expulsion of urine is affected by the contraction of the muscular fibres in the wall of the bladder—the detrusor urinæ—aided by the contraction of the voluntary muscles in the abdominal wall.¹ According to Goltz, the contraction of the detrusor is a purely reflex act. The reflex centre, which is situated in the segments of the spinal cord corresponding to the 3d, 4th, and 5th sacral nerves, is set into action by stimulation of the sensory fibres in the mucous membrane of the bladder.

Many authorities believe that the reflex process can be strengthened, and some say that it can be set into motion by a voluntary effort. Professors A. Mosso and P. Pellacani, conclude, as the result of elaborate and careful experiments on the human subject, ‘that *every psychological act*, intellectual, emotional, volitional, as well as every sensory perception, is accompanied by a contraction of the bladder.’ Moreover, they prove that the bladder can be made to contract by a pure act of the will apart from any sensory reflex, without the concomitance of abdominal pressure.² Like many other reflex acts, it can be inhibited by an effort of the will.

¹ Possibly the tonic contraction of the sphincter may be due to some local nervous mechanism.

² *Sur les fonctions de la Vessie.* By Prof. A. Mosso and P. Pellacani, quoted in *Brain*, July 1882, p. 239.

Some authorities conclude that the centres for the detrusor and sphincter muscles are antagonistic, and that an impulse from the brain, which inhibits the action of the sphincter centre, of necessity excites the action of the detrusor; and *vice versa*. Whether this is so in reality is doubtful. According to Professors Mosso and Pellacani there is no antagonism between them, every contraction of the one being accompanied by a contraction of the other; according to these observers it is only when the contraction of the detrusor becomes sufficiently powerful to overcome the resistance of the sphincter that the expulsion of urine is effected. The inhibitory fibres probably pass down the lateral columns of the cord.

The parts concerned in micturition, while at rest, are diagrammatically represented in fig. 144.

The mechanism of micturition then, so far as it is at present known, would seem to be as follows:—

1. When the bladder becomes sufficiently full of water, or rather, when the pressure on the interior of its walls is raised to a certain height, the sensory nerve filaments in the mucous membrane are stimulated, and an impression is conveyed along the sensory nerves to the reflex centres for the detrusor and sphincter muscles in the spinal cord, and to the sensorium.

2. As a result of the sensory impression conveyed to the brain the desire to urinate is experienced.

3. As a result of the reflex impulse carried to the spinal cord, the action of the detrusor centre is stimulated, while the action of the sphincter centre is inhibited.

If the circumstances for urination are favourable, an impulse is sent from the brain to: (a) the tonic centre for the sphincter, inhibiting its action, and causing relaxation of the sphincter muscle; (b) the centre for the detrusor, strengthening its action which has been already excited by the reflex impulse from the bladder; and to (c) the abdominal muscles, causing their active contraction. In health all these processes are simultaneously accomplished, and urination is the result. The mechanism of the process is diagrammatically represented in fig. 145.

If the circumstances are not convenient for urination, the process can be prevented by: (a) voluntary inhibition of the centre for the detrusor; (b) the (voluntary) contraction of the urethral muscles at the neck of the bladder; and possibly, too, the strengthening of the tonic centre for the sphincter. The manner in which the process is inhibited by a voluntary effort, is diagrammatically represented in fig. 146.

Alterations of the process of urination which occur in disease.—Disorders of urination are very frequent in disease, and result

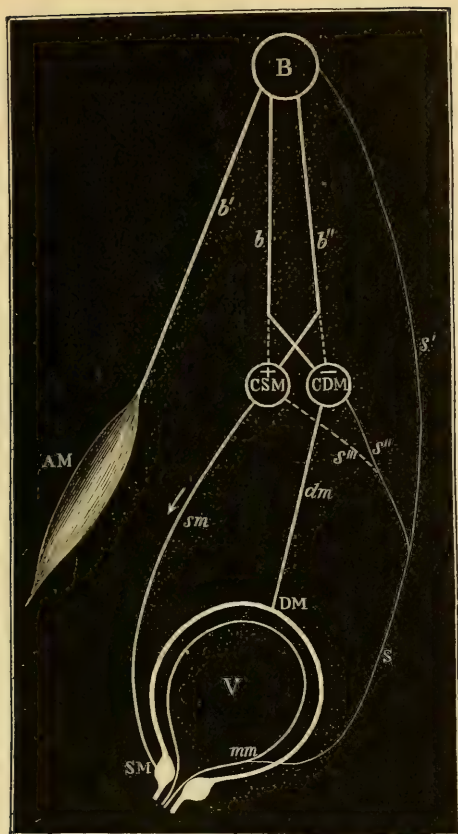


FIG. 144.—*Diagrammatic representation of the parts concerned in the mechanism of micturition while at rest.*—(After Gowers, but considerably modified.)

The sphincter muscle (S M) is in a state of contraction, the result of nerve force continually sent to it from its tonic centre (C S M) in the spinal cord, through *sm*, as indicated by the arrow.

V = the bladder, which is represented as empty. S M, sphincter muscle. D M, detrusor muscle. A M, abdominal muscles. *mm*, mucous membrane of bladder. B, the brain. C D M, spinal centre for the detrusor muscle. C S M, spinal centre for the sphincter muscle. S, sensory fibre proceeding from the mucous membrane of bladder up to the spinal cord and brain. *dm*, motor nerve from the spinal centre for the detrusor muscle. *sm*, motor nerve from the spinal centre for the sphincter muscle. *b*, nerve filament proceeding from the brain to the spinal centres of the detrusor and sphincter muscles. An impulse from the brain through *b* inhibits the sphincter centre (dotted line) and excites the detrusor centre, as shown in fig. 146. *b'*, Nerve filament proceeding from the brain to the spinal centres of the sphincter and detrusor muscles. An impulse along *b'* strengthens the sphincter and inhibits the detrusor centre. *b''* Nerve filament from the brain to the abdominal muscles.

Note.—When the bladder is empty its walls are collapsed. The condition represented in the figure is purely diagrammatic.

from derangement of the mechanism which I have just described; and since the chief part of that mechanism is situated in the

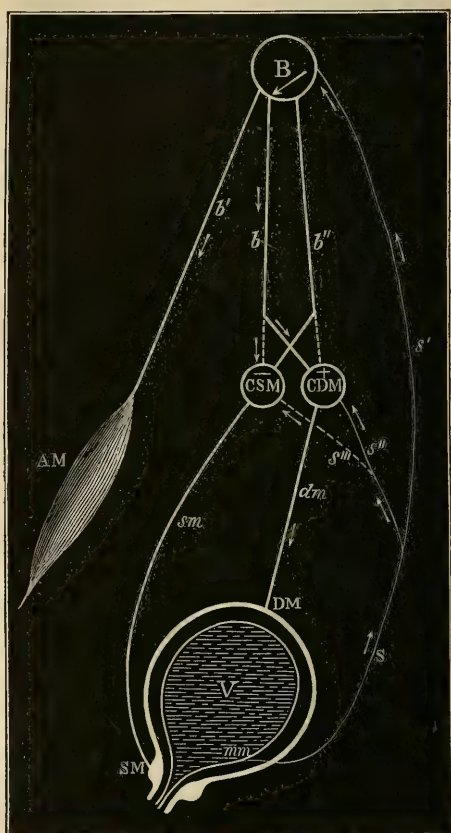


FIG. 145.—*Diagrammatic representation of the parts concerned in the process of micturition while in action.*

The bladder (V) is full of urine. The mucous membrane (mm) is stimulated, an impression is generated, and is carried by the sensory (centripetal) nerve (S) to the brain (B), and to the spinal centres for the detrusor (C D M), and sphincter (C S M). From the brain an impulse is sent (1) along the nerve (b) which strengthens the action of the detrusor centre (C D M), and inhibits the action of the sphincter centre (C S M); (2) along the nerve (b'), which throws the abdominal muscles (A M) into action.

The reflex impulse which has passed from the mucous membrane of the bladder to the spinal cord, excites the action of the detrusor centre, and inhibits the action of the sphincter centre.

The final result is contraction of the detrusor muscle (D M), relaxation of the sphincter muscle (S M); and the expulsion of urine. The arrows indicate the direction of the nerve 'currents.'

lower portion of the spinal cord, it follows that the greatest disturbance in urination will occur when the lumbar portion of the cord is the seat of the lesion.

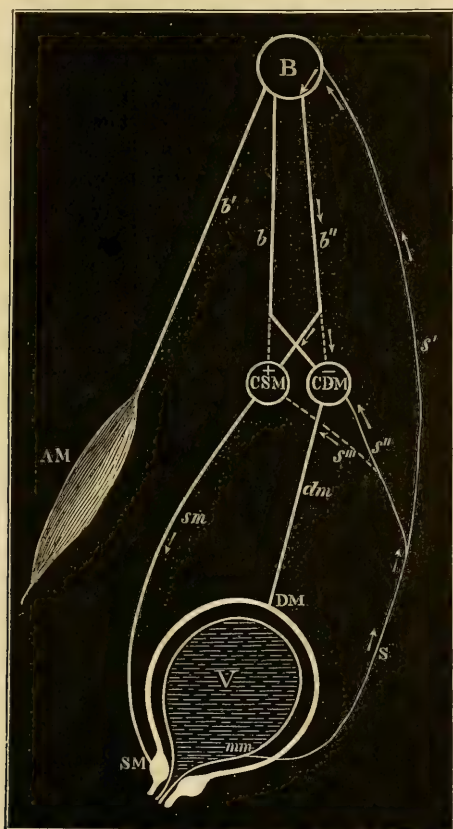


FIG. 146.—*Diagrammatic representation of the mechanism of micturition, showing the process by which the act can be inhibited by a voluntary effort.*

The bladder is full of urine. A reflex impulse passes from the mucous membrane to the spinal cord and brain; but the conditions for urination are not favourable. An impulse is therefore sent from the brain (B) along the nerve (b''), which inhibits the action of the detrusor centre (C D M), and strengthens the action of the sphincter centre (C S M). The final result being firm contraction of the sphincter muscle (S M), and *non*-expulsion of urine. The arrows indicate the direction of the nerve 'currents.'

The following are the effects produced by lesions of the different parts of this nervous mechanism:—

1. *Excessive stimulation of the centripetal nerves.*—When the sensory nerve filaments in the mucous membrane of the bladder

are abnormally sensitive, as in cases of cystitis, a small quantity of urine will suffice to set up the reflex process; spasmodic contractions of the detrusor muscle and increased frequency of micturition result. A foreign body (such as a stone) will produce excessive stimulation of the sensory nerves, even when the mucous membrane is healthy, but as a matter of fact cystitis is nearly always present in such cases. Irritation in the rectum may also excite the reflex mechanism. Some cases of nocturnal incontinence in children are probably induced in this manner.

2. *Destruction of the sensory (centripetal) nerve filaments* will, of course, prevent the reflex impulse passing to the spinal cord. Expulsion of urine can still, to some extent, be effected by voluntary effort, chiefly by means of the abdominal muscles. This condition is more of theoretical than practical importance. It might result from the pressure of a tumour or inflammatory products on the nerve trunk or posterior nerve roots.

3. *Destruction of the reflex centres in the spinal cord.*—Destruction of the reflex centres may result from myelitis, hæmorrhage into the cord, traumatic injuries, etc. The sphincter and detrusor muscles are, of course, paralysed. There is generally paraplegia and paralysis of the rectum.

Sudden injuries (traumatic and inflammatory, etc.) of the spinal cord above the lumbar region are also attended with arrested functions of the urinary centres. In cases of this description, the urinary reflex is re-established as soon as the effects of shock pass off (provided, of course, the lumbar cord remains unaffected). In many of these cases the conducting paths to and from the brain are interrupted with the results described below.

Destruction of the detrusor centre produces paralysis of the detrusor muscle, as a result of which urine collects in the bladder (retention): after a time the sphincter gives way, and incontinence occurs.

When the action of the detrusor centre is impaired but not destroyed, the paralysis is incomplete. In these cases the contraction of the abdominal muscles takes a larger share in the process than in health. The expulsive power is diminished; the patient cannot empty the bladder when lying on his back; in order to complete the process satisfactorily, he has to stand up, i.e. to place the parts in the best hydrostatic condition for the exit of urine.

Paralysis of the detrusor from destruction of its reflex centre is almost invariably associated with paralysis of the sphincter.

Destruction of the sphincter centre produces paralysis of the sphincter; the urine dribbles away; this condition is termed incontinence of urine. When the sphincter is paralysed, any

sudden movement on the part of the patient—laughing, coughing, etc.—forces away urine.

Paralysis of the sphincter is very rare *per se*. It is nearly always associated with a similar affection of the detrusor muscle and with paralysis of the rectum.

4. *Destruction of the centrifugal (motor) nerves*.—Destruction of the motor nerve which supplies the detrusor will, of course, produce paralysis of that muscle. As I have previously remarked, this condition is seldom, if ever, met with in practice.

5. *Interruption and destruction of the conducting paths to and from the brain above the reflex centre* are of frequent occurrence. If the lesion is a sudden one, the shock to the urinary centres may cause temporary arrest of function. In chronic cases the effects of the lesion vary with its position and extent. When the sensory conductors or sensory perceptive centres are affected, the desire to urinate is not perceived; the reflex arc is uninjured, and as soon as the bladder becomes sufficiently distended with urine, it is evacuated unconsciously. It must be remembered, therefore, that the involuntary discharge of urine and fæces in cases of coma does not necessarily imply any paralysis of the bladder or rectum.

When the motor and inhibitory fibres are alone interfered with, the desire to urinate is perceived, but the act takes place quite independently of volition. It can neither be assisted nor prevented.

The remarks which I have just made with regard to the bladder reflex, apply to the rectal reflex. Obstinate constipation is a conspicuous feature in many cases of severe myelitis. When the myelitis involves and destroys the reflex centre for the rectum, the sphincter is paralysed and there is incontinence of fæces. When the lesion is situated higher up, there is obstinate constipation, or (but this is uncommon) ‘unconscious reflex defæcation.’

These differences in the nature and severity of the bladder paralysis are not always so clearly marked at the bedside as the statements which I have just made might lead you to suppose. The important point is that complete and permanent paralysis of the sphincters rarely if ever results except in those cases in which the reflex centres for the bladder and rectum are directly involved, i.e. in which the myelitis involves in the first instance, or subsequently extends to, the lower part of the cord. But grave paralysis of the bladder and serious

bladder complications are frequently the result of a myelitis in the upper lumbar and lower dorsal regions.

And this leads me to say that an ammoniacal condition of the urine and septic cystitis are of frequent occurrence in cases of severe myelitis; they are a source of great danger to the patient. In some cases, the cystitis and ammoniacal condition of the urine are due to the introduction of septic germs into the bladder from without. This form of cystitis may be developed either in the earlier or the later stages of the case. I cannot too forcibly impress upon you the importance of cleanliness and of the strictest antiseptic precautions, when catheterisation is required. In other cases—and in these cases the myelitis is always severe—the urine becomes ammoniacal and septic cystitis is developed in the early stages of the case, it may be within a few days from the commencement of the attack, apparently independently of the introduction of any septic spark from the outside. This grave form of acute septic cystitis attended with ammoniacal urine, blood, pus and mucus in the urine, and often associated, as has been determined after death, by ulceration of the bladder walls, may be developed in cases in which the sphincter is paralysed and in which the catheter has not been required. It is supposed to be the result of a trophic lesion in the walls of the bladder exactly analogous to another trophic lesion which I shall presently mention, namely, the acute trophic bed-sore.

Septic cystitis and an ammoniacal condition of the urine may be developed during the stage of spastic paraplegia which follows the myelitis, or, if it has been developed during the acute stage of the disease, may continue for months or even years.

Many paraplegic patients, who have recovered sufficiently to walk about and even to follow their business occupations, die from cystitis and the kidney lesion which results from it, several years after the original attack. In more than one case of this kind, I have found the walls of the bladder enormously thickened, and covered with a thick phosphatic crust.

Bed-sores.—I have said that the acute form of cystitis is comparable to the acute form of bed-sore; and this leads me to refer to bed-sores. They are frequently developed in cases of transverse myelitis. There are two forms; both are of the greatest practical importance.

The common variety is the result of continuous pressure and irritation of the skin the vitality and resisting power of which are lowered by the cord lesion. In consequence of the paralysis, the patient lies like a log in one position, unable to move; in many cases, unable to feel; and, unless he is very carefully nursed, often wet and dirty. It is no wonder that under such circumstances bed-sores often form on the back,

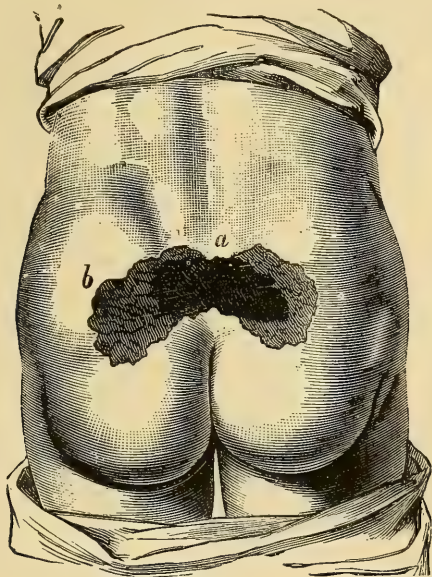


FIG. 147.—*The acute bed-sore, from a case of myelitis, which involved the dorsal region of the cord.*—(After Charcot.)

a, slough; *b*, erythematous zone.

hips and other parts which are subjected to continuous pressure and external irritation. When I come to speak of the treatment, I shall have to tell you that one of the most important points in the treatment of cases of acute myelitis is the prevention of cystitis and of bed-sores. This point cannot be too frequently repeated or too forcibly emphasised.

The other form of bed-sore, which is fortunately less common, is termed the acute bed-sore (see fig. 147). It is due to a gangrenous inflammation of the skin, which develops in the

early stage of the case, it may be within a few hours or days from the commencement of the attack. The acute bed-sore is, in some cases, a true trophic lesion, for it may form on parts of the skin which have not been subjected to continuous pressure or irritation. But although this may undoubtedly occur, it is certain, I think, that in most cases the acute bed-sore is partly due to pressure and external irritation. The hips, back, inner sides of the knees, inner sides of the ankles and the heels are the parts on which acute bed-sores are usually developed. It is often difficult or impossible to prevent the formation of the acute form of bed-sore; for the trophic



FIG. 148.—*Trophic lesions in the toes, the result of a severe acute myelitis.*

disturbance in the nutrition of the skin is so great that a slight degree of external pressure or irritation is sufficient to excite a gangrenous inflammation.

In cases of severe myelitis, trophic alterations in the skin, nails and hair may occur. Bullæ and ulcers may form; the skin may become smooth and glossy; the nails may become brittle and cracked or become detached; the ends of the toes may ulcerate and drop off (see fig. 148).

The acute bed-sore and the acute form of cystitis are most frequently developed in those cases in which the inflammation involves the grey matter of the lumbar and sacral segments, especially perhaps the grey matter of the posterior horn.

Septic cystitis and bed-sores are apt to be attended with

fever of a septic or pyæmic type and with profound constitutional disturbance. Acute septic cystitis and acute bed-sores often prove fatal.

Before concluding the description of the clinical history, let me briefly refer to some other symptoms which are occasionally developed in the course of the disease.

Girdle sensation.—In some cases of myelitis, the patient complains of a feeling of constriction, as if a cord were tied tightly round the waist. To this condition the term 'girdle sensation' has been applied; it is often associated with, and due to, a localised band of hyperæsthesia. The band of hyperæsthesia is of importance from a diagnostic point of view, for it marks the upper limit of the lesion. It is due to the irritation of the sensory fibres of the posterior roots at the upper end of the lesion, i.e. at the part of the cord where the inflammation abuts upon healthy cord tissue.

Paralysis of the respiratory muscles.—When the myelitis involves the upper dorsal region, the thoracic muscles may be paralysed. This is a very serious complication. In the very rare cases in which the myelitis affects the upper cervical region, the diaphragm may also be involved. A myelitis which ascends and produces paralysis of the respiratory muscles and of the diaphragm soon proves fatal.

Hyperpyrexia is developed in some cases, more especially when the inflammation involves or extends to the cervical region or the medulla oblongata.

Priapism is developed in some cases of myelitis, especially in those cases in which the lesion is situated in the upper dorsal or cervical regions. When the inflammation involves the lumbar and sacral segments, the penis is flaccid.

Alterations in the size of the pupils are sometimes observed when the cilio-spinal region of the cord is the seat of the inflammation.

Optic neuritis has also been noted in a few cases of myelitis, and especially in the disseminated form. Its exact mode of production is obscure. In all probability it is the result of a common toxic cause acting upon the spinal cord and optic nerve, rather than the direct result of the cord lesion.

Summary of the symptoms in cases of total transverse myelitis.—The more important symptoms which result from acute destruc-

tion of the whole transverse section of one segment are diagrammatically represented in fig. 149; they are as follows:—

1. Paralysis of (a) the muscular area of the affected segment and of (b) all the muscular areas which are situated below it.

2. Rapid atrophy, the 'reaction of degeneration' and complete absence of all reflex movements in the muscular area of the affected segment.

3. The absence of rapid atrophy and the reaction of degeneration in the muscles innervated by segments below the lesion.

4. Exaggeration of the reflexes passing through the segments below the lesion, and tension and rigidity of the paralysed

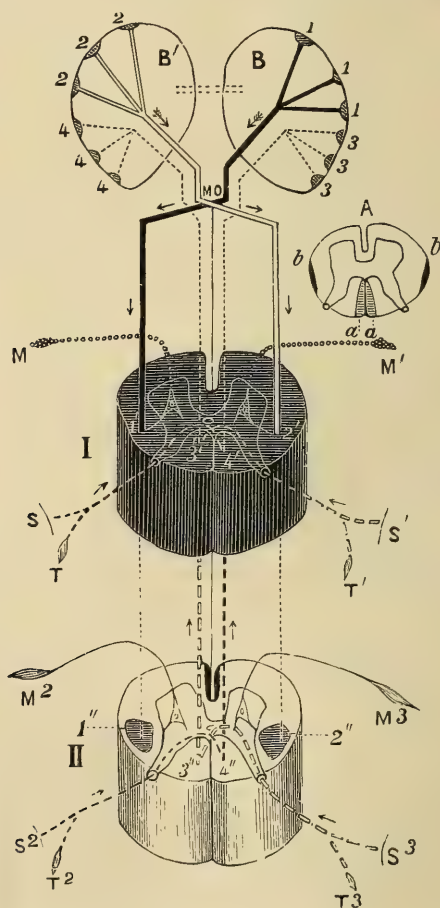


FIG. 149.

muscles innervated by inferior segments. These symptoms are developed after the initial effects of the lesion pass off and when secondary descending degeneration is developed. As I have previously pointed out, Dr. Charlton Bastian thinks that in those cases in which the whole thickness of the cord is completely destroyed and the connection between the parts below the lesion and the brain *completely* interrupted, the reflexes remain abolished in the parts below the lesion.

5. Diminution or abolition of the common and special sensibility (muscular sense) of the paralysed muscles.

6. Complete anæsthesia of the sensitive area of the affected segment and of the sensitive areas of all the segments which are situated below it.

7. The presence of a narrow band of hyperæsthesia at the upper level of the lesion, which is supposed to be due to irritation of the sensory nerve fibres at the upper level of the lesion. When the lesion is situated in the dorsal region of the cord, the band of hyperæsthesia extends round the trunk in the form of a

DESCRIPTION OF FIG. 149.

Diagrammatic representation of the effects of an acute total transverse lesion of the spinal cord.

I. The segment which is destroyed by the lesion ; II. A segment of the cord below the lesion ; B, the right, and B' the left cerebral hemispheres ; 1, 1, 1, motor centres in the right cerebral hemisphere from which the motor tract proceeds to the muscles M and M² on the left side of the body. (For the sake of clearness, the direct pyramidal tracts have been omitted.) 2, 2, 2, motor centres in the left cerebral hemisphere from which the motor tract proceeds to the muscles M¹ and M³ on the right side of the body ; 3, 3, 3, sensory centres in the right cerebral hemisphere to which the sensory tract proceeds from S, T, and S², T², the sensory areas on the left side of the body ; 4, 4, 4, sensory centres in the left cerebral hemisphere to which the sensory tract proceeds from S', T' and S'³, T'³, the sensory areas on the right side of the body.

Motor impulses passing downwards through 1 and 2 are arrested at the seat of the lesion. The muscles M and M' supplied by the diseased segment are in a state of acute atrophy. The muscles M² and M³ supplied by segment II. retain their normal bulk. Reflex impulses cannot pass through the diseased segment. Reflex impulses passing through segment II. are exaggerated, for the reflex arc is perfect, and the lateral columns above this segment are degenerated. Sensory impressions passing from all the parts below the lesion are arrested in the diseased segment. Secondary descending degeneration of the crossed and direct pyramidal tracts below the lesion, is indicated by the dotted lines and is shown in segment II. Secondary ascending degeneration above the lesion is indicated by the dotted continuations of the sensory tracts and is shown at *a, a*, in fig. A. For the sake of clearness, secondary ascending degeneration of the direct cerebellar tracts is not shown in the large figure ; it is, however, seen (*b, b*), in fig. A.

Note to Figs. 149 and 151.

In these figures (1) a total sensory decussation in the cord is shown, and (2) the sensory fibres are made to pass up the postero-internal column.

belt. The patient feels as if a tight band were drawn round his body; hence the term, 'girdle sensation.' When the lesion involves the lumbar or cervical segments, the band of hyperæsthesia, instead of being circular, is distributed more or less longitudinally over the limbs in accordance with the particular segment which happens to be affected. In fig. 150, I have

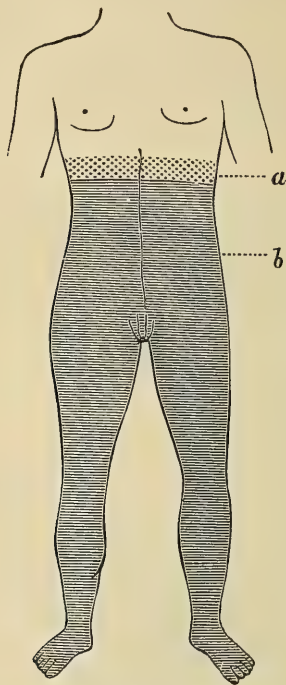


FIG. 150.—*Diagrammatic representation of the skin symptoms in a total transverse lesion of the dorsal portion of the spinal cord.*

The horizontal shading signifies anæsthesia of the skin; the dotted shading hyperæsthesia; *a*, hyperæsthetic zone at the upper level of the lesion; *b*, anæsthetic below the lesion.

diagrammatically represented the skin symptoms which result from a total transverse lesion.

8. Some vasomotor paralysis below the lesion.

9. Trophic disturbances in the skin of the sensitive area of the segment, and in many cases bed-sores.

10. Paralysis of the bladder and rectum, and in many cases ammoniacal urine and cystitis. The exact character of the vesical and rectal derangement, which depends upon the

condition of the reflex centres for the bladder and rectum, has been fully described above.

It should further be noted that secondary descending degeneration of the pyramidal tracts (crossed and direct), below the lesion; and secondary ascending degeneration of the postero-internal columns of Gowers' ascending tract and (if the lesion is situated in the dorsal or cervical regions) of the direct cerebellar tracts, above the lesion, are gradually established.

Summary of the symptoms in cases of a unilateral transverse myelitis.—The more important symptoms which follow acute destruction of one-half of a segment of the spinal cord are diagrammatically represented in fig. 151; they are as follows:—

1. Paralysis of (a) the muscular area supplied by the anterior root arising from the affected half segment; and (b) of all the muscular areas on the same side of the body which are situated below it.

2. Flaccidity, rapid atrophy and the 'reaction of degeneration' in the paralysed muscles supplied by the affected half segment.

3. The absence of rapid atrophy and the 'reaction of degeneration' in the paralysed muscles supplied by half segments below the lesion.

4. With the occurrence of secondary descending degeneration, the muscles supplied by half segments below the lesion become tense and rigid.

5. Certain reflex alterations. Theoretically we should expect to find that the reflexes passing through the affected half segment would become abolished; and that the reflexes passing through the inferior half segments on the same side of the body would become increased. But the exact condition of the reflexes in unilateral lesions has not yet been sufficiently investigated to allow of any very positive statement being made.

6. Diminution or abolition of the common and special sensibility (muscular sense) of the paralysed muscles.

7. Vasomotor paralysis below the lesion on the same side.

8. Anæsthesia of the sensitive area of the affected segment. The anæsthesia is bilateral; for the sensory fibres which enter by the posterior nerve root of the affected side, are 'cut' before their decussation, and the sensory fibres which enter the segment by the posterior root of the opposite side, are 'cut' after they have decussated. When the lesion is situated in the dorsal region, the band of anæsthesia is circular (see fig. 152); where the cervical or lumbar segments are involved, it is, for the

reasons already given in speaking of total transverse lesions, more or less longitudinal.

9. Anæsthesia of the sensitive areas of all inferior half segments on the opposite side of the body.

10. Hyperæsthesia of the sensitive areas of all inferior segments on the same side of the body. The exact cause of the hyperæsthesia is not known.

11. A narrow band of hyperæsthesia corresponding to the upper level of the lesion on the same side of the body. It is supposed to be caused by irritation of the sensory fibres which enter the cord at the upper level of the lesion. The band of hyperæsthesia is circular in the cases in which the lesion is situated in the dorsal region (see fig. 152); and more or less

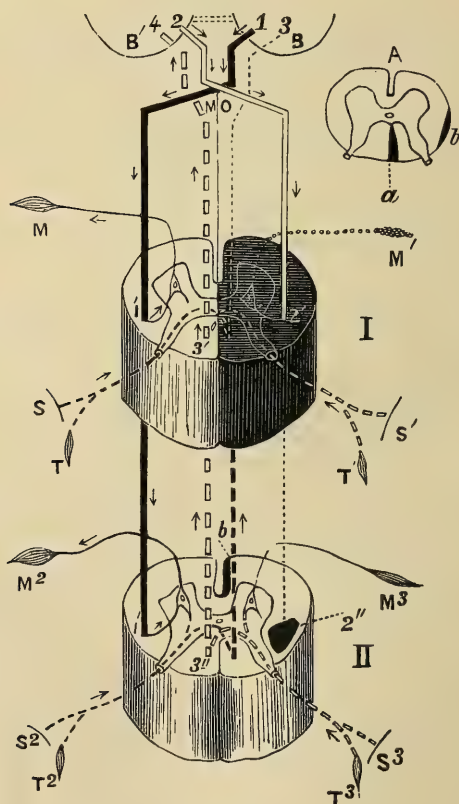


FIG. 151.

longitudinal in those cases in which the lesion is situated in the cervical or lumbar segments.

The character and distribution of the sensory disturbances in cases of unilateral lesion are well shown in fig. 152.

12. Derangements of the vesical and rectal reflexes may occur.

13. Trophic disturbances in the skin of the sensitive area of the affected half segment are occasionally observed.

Secondary degenerations.—Secondary descending degeneration of the pyramidal (crossed and direct) tracts on the same side below the lesion, and secondary ascending degeneration of the postero-internal column, of Gowers' ascending tract and of the direct cerebellar tract on the same side above the lesion, will of course follow.

The fibres of the posterior root which enter the affected half segment and their upward prolongations in the opposite postero-internal column will also degenerate; but the fibres entering by any one posterior root form such a small proportion of the whole sensory tract that this ascending degeneration on the opposite side to the lesion may not be perceptible.

The Modes of Termination of the Acute Stage and the Subsequent Course of Cases of Acute Transverse Myelitis.—The acute stage not unfrequently terminates in death; the fatal

DESCRIPTION OF FIG. 151.

Diagrammatic representation of the effects of an acute unilateral transverse lesion of the spinal cord.

I. A segment of the spinal cord, the right half of which is destroyed by the lesion; II. a segment of the spinal cord below the lesion; 1, motor tract proceeding from the right hemisphere of the brain B, to the muscles M, and M², on the left side of the body (for the sake of clearness the direct pyramidal tracts have been omitted); 2, motor tract proceeding from the left hemisphere of the brain B', to the muscles M¹ and M³, on the right side of the body; 3, the sensory tract proceeding to the right hemisphere of the brain from S, T, and from S², T², the sensitive areas on the left side of the body; 4, the sensory tract proceeding to the left hemisphere of the brain from S¹, T¹, and from S³, T³, the sensitive areas on the right side of the body.

Motor impulses, passing from the left hemisphere of the brain along 2, are arrested at the seat of the lesion. There is, therefore, paralysis of the muscular areas M¹ and M³. The motor tract below 2' is degenerated. The muscular area M¹ is acutely atrophied. Reflex impulses, passing from S¹, T¹ to M¹, are abolished. The paralysed muscles M³ below the lesion are not atrophied. Reflexes passing from S³ and T³ to M³, are exaggerated, for the reflex arc which passes through segment II. is intact, and the lateral column above segment II. is degenerated; the degeneration of the motor tract below 2' is indicated by a dotted line, and is seen in segment II. Sensory impressions from the sensitive areas, below the lesion, and on the opposite side of the body, S² T², are arrested at the seat of the lesion. Sensory impressions from S, T, and S¹, T¹, the sensitive area of the affected segment on both sides of the body, are also 'blocked' by the lesion.

result may be due to a variety of causes, amongst which the following are some of the chief:—paralysis of the respiratory

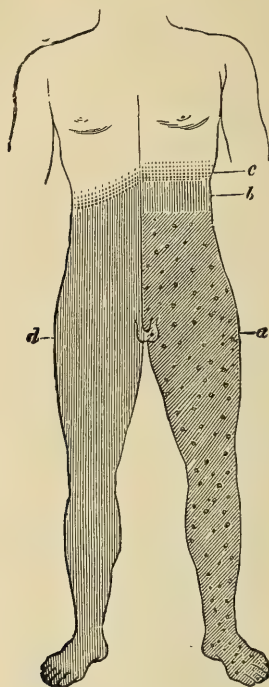


FIG. 152.—*Diagrammatic representation of the skin symptoms in a unilateral lesion of the dorsal portion of the spinal cord on the left side.*—(After Erb.)

The diagonal shading signifies motor and vasomotor paralysis; the vertical shading signifies anæsthesia of the skin; the dotted shading indicates hyperæsthesia of the skin; *c*, band of hyperæsthesia on the left side of the body at the upper level of the lesion; *b*, band of anæsthesia on the left side of the body corresponding to the vertical extent of the lesion; *a*, hyperæsthesia below the level of the lesion; *d*, anæsthesia, in the parts below the level of the lesion on the opposite (right) side of the body.

muscles; cystitis and kidney complications; bed-sores and blood poisoning; pneumonia or other pulmonary affections.

In most cases of transverse myelitis, the acute stage is tided over, the condition becomes chronic, and a stage of cicatrization and secondary degeneration is developed. The future course of events varies greatly in different cases.

After the acute symptoms subside, the general health is

gradually re-established and a condition of chronic paraplegia remains. In many cases, a considerable amount of motor power is slowly regained and sensation in the paralysed parts is more or less completely restored. Finally, the patient may be able to walk about with the aid of crutches or sticks. In a few cases, the recovery is much more complete; and occasionally, but very rarely, a perfect cure is established. As the process of cicatrisation and secondary descending degeneration advances, the paralysed muscles, below the level of the lesion, become rigid; their reflexes (especially the deep reflexes) become exaggerated; and the paraplegia assumes the spastic type. In short, as I have more than once pointed out, acute and chronic myelitis are the usual causes of spastic paraplegia.

In other cases, little or no improvement occurs. After the acute symptoms have subsided, the patient may remain for some time *in statu quo*. Ultimately, he may die, exhausted by the discharge from extensive bed-sores or in consequence of the development of kidney or respiratory complications.

A severe myelitis of the lumbar enlargement is rarely if ever completely recovered from. The patient remains paraplegic; the paralysed parts, however, instead of becoming spastic and rigid may remain flaccid, some of the muscles being markedly wasted; this is a much more serious and hopeless form of paraplegia than the spastic variety, but it is fortunately rare.

Let me now briefly refer to some other varieties of the disease.

Central Myelitis.—This is a very grave form of the disease; the central grey matter is chiefly implicated; but the white columns may also of course be involved, though rarely in the same degree. The inflammation is apt to extend and to involve an extensive area of the cord. In those cases in which the inflammation pursues an ascending course, death may occur in the course of a few days from involvement of the respiratory muscles. Motor paralysis, extensive in distribution and rapidly progressive, loss of sensation and especially of the sensibility to pain and to heat and cold, and trophic disturbances are the most marked symptoms in this form of myelitis, together with fever, which is often high, and profound constitutional disturbance.

Ascending Myelitis.—In this variety, the symptoms are very similar to those which have just been described as characteristic of central myelitis. Like central myelitis, this is a very grave form; it usually proves fatal in the course of a few days in consequence of respiratory paralysis or extension of the inflammation to the medulla oblongata.

Disseminated Myelitis.—In this variety, the inflammatory process is more frequently subacute than acute. The clinical picture may be a very complicated one; for a number of different foci of inflammation may be developed one after the other in different parts of the cord. Acute disseminated myelitis is probably always due to a toxic cause; it usually follows an acute febrile disease, such as measles, small-pox, pyæmia, gonorrhœa, erysipelas, etc. In the subacute forms, syphilis is I believe by far the most common cause. The inflammatory foci usually develop, as Dreschfeld has pointed out, around the blood-vessels. Optic neuritis has been observed in several cases. In Dreschfeld's interesting case (reported in the *British Medical Journal*, June 2nd, 1894), the optic neuritis developed before the cord symptoms. As in the central and ascending forms of myelitis, death is often due to paralysis of the respiratory muscles.

Localised or Focal Myelitis.—In this variety, the symptoms are much less conspicuous. Indeed, in those cases in which the inflammatory focus is very minute the symptoms may be so trivial as to escape detection; but cases of this kind are by no means unimportant, for a minute focus of inflammation may be the starting-point of a more extensive chronic lesion. It is probable, I think, that in some cases in which spastic paraplegia appears to be primary, the lesion in the crossed pyramidal tracts has its starting-point in a localised myelitis. Further, acute transverse myelitis is sometimes developed around a localised inflammatory focus. I have met with more than one case in which, during convalescence from a slight and localised myelitis, an acute exacerbation followed by complete paraplegia occurred as the result of exposure to cold, violent exercise, or some other exciting cause.

Compression Myelitis.—This form of myelitis, which is rarely acute except as the result of traumatic injury to the spinal bones

(fracture or dislocation of the vertebræ), will be more appropriately considered after I have described the important subject of inflammation of the spinal membranes (see page 570).

We have now considered all the more important points in the clinical history of acute transverse myelitis. In the lecture to-morrow I propose to direct your attention to the diagnosis, prognosis and treatment of the disease.

LECTURE XXVII

ACUTE MYELITIS (*Continued*)

LET us now turn, Gentlemen, to the diagnosis and differential diagnosis.

Diagnosis.—In well-marked and typical cases, the diagnosis of acute transverse myelitis is not attended with difficulty. The more important features from a diagnostic point of view are:—The quickly developed paraplegia, the fever and constitutional disturbance (though the fever and constitutional disturbance are, as we have seen, in some cases slight or absent); the loss of sensation in the paralysed parts, the paralysis of the bladder and rectum, and the bed-sores which in many cases are developed.

When these symptoms occur in a previously healthy individual, when they are limited to the lower parts of the body and when they do not show any tendency to extend upwards, acute transverse myelitis may be diagnosed, provided that primary hæmorrhage can be excluded.

It is in the less severe forms of acute transverse myelitis, and in the focal, disseminated and central forms of myelitis that the chief difficulties in diagnosis occur.

The differential diagnosis of acute transverse myelitis and of poliomyelitis anterior acuta.—A transverse myelitis is at once distinguished from poliomyelitis anterior acuta by the profound disturbance of sensation (anæsthesia), the marked paralysis of the bladder and rectum, and the bed-sores which are so frequently developed in severe cases.

The distinction is specially easy in those cases in which the dorsal region is affected, for in these cases the muscles of the lower extremities do not undergo the rapid form of atrophy and do not present the reaction of degeneration, and the deep

reflexes, which may be abolished in the early stages, are after a little time exaggerated.¹

An indiscriminate myelitis which happened to be limited to the region of the anterior cornua could not be distinguished from the system disease (poliomyelitis anterior acuta); the symptoms in the two cases would be absolutely the same. It is probable that an indiscriminate myelitis is rarely limited in this way, but it is not unlikely that some of the cases which appear to be cases of poliomyelitis anterior acuta in the adult are in reality due to an indiscriminate myelitis which is accidentally limited to the anterior cornual region.

The differential diagnosis of myelitis and spinal meningitis.—Myelitis is easily distinguished from spinal meningitis. You will appreciate the points of distinction better after I have described spinal meningitis; but I may say that spinal meningitis is characterised at its commencement by a prolonged stage of irritation in which pain in the back, pain in the limbs often distributed in the course of special nerve roots, cramps and spasms in the muscles and rigidity of the spine are prominent symptoms; whereas in myelitis the irritative phenomena are rarely prominent and are always of short duration. Further, in meningitis, motor paralysis (paraplegia), if developed at all, is rarely developed during the early stages of the attack; and paralysis of the bladder and rectum are much less prominent symptoms than in myelitis, though retention of urine is not uncommon. Again, in cases of meningitis, bed-sores are never developed in the early stages of the case. You must, however, remember that meningitis and myelitis are sometimes combined. But in cases of meningo-myelitis there is usually no difficulty in saying whether the meningitis or the myelitis is the predominant lesion. In many of the cases in which well-marked symptoms of myelitis are associated with well-marked symptoms of meningitis, the inflammation of the spinal cord and spinal membranes is due to a common cause—a well-marked extramedullary lesion for example, such as disease of the bones, fracture or dislocation of the spine, etc.

The differential diagnosis of myelitis and functional paraplegia.—The diagnosis of paraplegia due to myelitis and functional paraplegia is in some cases, more especially in those cases in

¹ See note on p. 127.

which the myelitis is subacute or chronic, attended with great difficulty. The same difficulty occurs in connection with many other diseases of the nervous system. The most experienced and skilled neurologists have sometimes to hesitate before giving a positive opinion. Further, it must never be forgotten that organic disease and functional disturbance are not unfrequently combined. So far as our present knowledge enables us to judge, functional paralyses are in the great majority of cases associated with, and apparently due to, the condition of the nervous system which we term hysterical. But it is a mistake to suppose that every functional paralysis is hysterical. Dr. Charlton Bastian, in a recent and most suggestive work, has laid special stress upon this point. Further, it is in my opinion a mistake to conclude that every case of paraplegia which is recovered from is functional. Complete recovery is by no means uncommon in cases in which the paralysis is undoubtedly due to organic disease. Personally I am disposed to think that some of the cases which (because they recover) are regarded as functional, are in reality due to organic disease.

In cases of functional paralysis, the seat of the functional disturbance is more frequently cerebral than spinal. Functional hemiplegia and functional monoplegia are for example more common than functional paraplegia; and in cases of functional paraplegia the primary functional derangement is more frequently situated in the cerebrum than in the spinal cord—a fact which must be kept in view in considering the differential diagnosis of paraplegia due to functional and organic conditions.

It is difficult to lay down any general rules which will enable the practitioner to determine in every case whether a paraplegia is due to organic disease or to functional derangement. Of course, in most cases the differential diagnosis is easily made; but individual cases present great differences in detail, and in the doubtful cases the practitioner has to rely upon his general knowledge of nervous diseases and the experience gained in dealing with previous cases of the same kind.

In doubtful cases, the diagnosis chiefly turns upon the condition of the paralysed muscles, the condition of the bladder and rectum, the presence or absence of bed-sores and the nature of the associated symptoms.

Bed-sores, especially if rapidly developed, paralysis of the sphincters and rapid atrophy with the reaction of degeneration are never, so far as I know, due to functional derangement; they only occur as the result of organic disease.

Complete abolition of the knee-jerks and a well-marked girdle sensation are highly suggestive of organic disease.

In some cases of functional paraplegia, the muscles of the lower extremities are flaccid and to a certain extent atrophied. In others, the lower limbs are in a condition of rigidity and tension.

In the flaccid cases, the atrophy is rarely great. In cases of this kind (functional paralysis with muscular flaccidity), the reaction of degeneration is, in my experience, never developed and the knee-jerks are never abolished; in fact, notwithstanding the flaccidity and atrophy, the knee-jerks are usually exaggerated.

In the spastic cases of functional paralysis, the muscular rigidity and tension and the exaggeration of the deep reflexes are seldom so marked as in spastic paraplegia the result of myelitis. In the functional cases, typical ankle-clonus is rarely, if ever, present. I used formerly to think that typical ankle-clonus was proof positive of organic disease. I now speak with more reserve upon this point; but I still hold that well-marked ankle-clonus is highly suggestive of organic disease. I have never myself seen typical and fully developed ankle-clonus in any case of functional or hysterical paralysis.

Retention of urine may of course occur in functional, more especially in hysterical, cases; but persisting (as distinguished from temporary and intermittent) paralysis of the bladder, and especially paralysis of the sphincter muscle, is in my experience indicative of organic disease. The acute bed-sore is absolutely indicative of organic disease.

In cases of functional paraplegia due to hysteria, the presence of associated symptoms and signs of the hysterical condition (such as emotional disturbances, convulsive seizures, patches of anaesthesia and hyperaesthesia, hemianæsthesia, the characteristic constriction of the fields of vision, ovarian tenderness, aphonia, etc.) are of great diagnostic significance.

In hysterical and functional cases, there are often localised tender areas over the spinal column.

The subjects of functional paraplegia are usually emaciated,

anæmic, or debilitated; in many cases, they have, prior to the onset of the paralysis, been subjected to overstrain or some exhausting or debilitating conditions.

The age and sex of the patient, and the mode of onset of the paralysis may afford corroborative evidence as to the true nature of the case. Functional paraplegia is much more common in women than in men, and is chiefly met with in young adult women and during the period of active sexual life. In some cases of functional paraplegia, the paralysis is suddenly developed after a convulsive seizure, fright, or other cause of emotional excitement.

A history of previous paralytic attacks which have been recovered from is highly suggestive of the functional character of the paralysis. I shall have more to say on this point when I come to speak of multiple cerebro-spinal sclerosis, a condition which is very apt in its early stages to be mistaken for hysterical paraplegia, and in which, as in some hysterical cases, remarkable temporary improvement in the paraplegic symptoms (in fact in some cases almost complete remissions) occasionally occur.

In those cases in which it is impossible by means of the foregoing considerations to come to a positive opinion, we must be content to observe the case and to watch the course of events, treating the paralysis as if we believed it to be curable. If we say to ourselves, 'This case is incurable,' we are not likely to make any very vigorous effort to cure it. As I have so repeatedly told you, hopefulness in dealing with disease is a very important quality; it is one of the first essentials for success in practice. But there are different forms of hopefulness. The man who looks hopefully upon a case of acute myelitis with bed-sores and paralysis of the sphincters, and who predicts that the paralysis will be recovered from, is not a reliable practitioner. I do not mean to say that such cases never recover. I have seen more than one such case in which recovery has been complete. But such a fortunate result is extremely rare. In the doubtful cases in which the practitioner is unable to decide whether the paralysis is due to organic disease or functional disturbance, the patient should have the benefit of the doubt; the treatment should be conducted on the supposition that the paraplegia is curable.

The differential diagnosis of primary and secondary myelitis.—When a diagnosis of myelitis has been arrived at, the further

question whether the inflammation of the spinal cord is primary or secondary, say, to changes in the bones or membranes, has to be determined. This question, which is of the greatest practical importance both for prognosis and treatment, chiefly arises in those cases in which the myelitis and the resulting paraplegia are sub-acute or chronic. Myelitis, due to disease of the bones and membranes (excepting, of course, the traumatic cases) is, comparatively speaking, rarely acute. I shall consider this point in more detail when I come to describe compression myelitis. I will then refer to several cases in which paraplegia was rapidly developed as the result of extra-medullary lesions (Pott's disease of the vertebræ, tumours of the vertebræ, etc.). In cases of this description, the condition of the spinal column and the fact whether the paraplegia was preceded by symptoms indicative of bone disease (pain in the back, etc.) or of pressure upon individual nerve roots are the points of special diagnostic importance.

The exact anatomical diagnosis of transverse myelitis.—The exact position of the inflammatory lesion can usually be determined without much difficulty by observing the distribution of the motor and sensory paralysis and the condition of the reflexes. In those cases in which a hyperæsthetic band is present, the upper limit of the lesion is clearly defined by the position of the band of hyperæsthesia. The vertical extent of the lesion is more difficult to determine.

The condition of the reflexes and the condition of the paralysed muscles as regards atrophy and the presence or absence of the reaction of degeneration, are our chief guides. We have seen that in cases of acute transverse myelitis, muscles supplied by motor nerve roots belonging to the affected segment or segments of the cord undergo rapid atrophy and present the reaction of degeneration, and that the reflexes passing through the affected segment or segments of the cord are abolished; while the muscles attached to segments below the lesion, although they may be completely paralysed, do not undergo rapid atrophy and do not present the reaction of degeneration; further, their reflexes (after the primary effects of shock have been recovered from) are exaggerated. In dealing, then, with a supposed case of transverse myelitis, the muscle-areas must be carefully examined from below upwards. If marked atrophy

and the reaction of degeneration are present in any of the paralysed muscles, we conclude that the muscles which exhibit these changes are connected with the segment or segments of the cord which are directly implicated by the lesion. This seems very simple in theory, but it is by no means so simple in practice. The lesion is often situated in the dorsal region and it is in many cases a difficult matter to determine whether the intercostal or abdominal muscles present these changes (rapid atrophy, etc.).

The condition of the abdominal reflex at different levels is much more readily determined, and is one of the simplest and best guides to the vertical extent of a transverse myelitis. When the lesion involves the dorsal region, the upper level of the anæsthesia and the exact position of the hyperæsthetic band can of course, be easily determined, for in such cases the anæsthesia and hyperæsthesia pass round the trunk in the form of a girdle. But it is more difficult to determine the upper level of the anæsthesia and to map out the exact distribution of the hyperæsthetic band in those cases in which the myelitis involves the lumbar and sacral portions of the cord; for in such cases the anæsthesia and hyperæsthesia are distributed more or less obliquely and longitudinally down the limbs in accordance with the distribution of the lumbar and sacral nerves.

○ **The differential diagnosis of acute transverse myelitis and of primary spinal hæmorrhage.**—I shall refer to this point when I come to describe spinal hæmorrhage. It is not perhaps of much importance from a practical point of view, for the treatment of primary hæmorrhage and of acute myelitis is very similar. Further, the distinction is not always possible, for, as I have already told you, localised hæmorrhages into the inflamed tissue are of frequent occurrence in cases of acute myelitis. In some cases of this kind, the hæmorrhage is so considerable and the paralysis is developed with such rapidity that a primary hæmorrhage is closely simulated. In cases of this description, the rapidity of onset (whether the paralysis is developed instantaneously or not) and the fact whether the suddenly developed paralysis is preceded by any symptoms indicative of commencing myelitis are the most important points for the purposes of diagnosis. When I come to describe spinal hæmorrhage, I shall tell you that in many of the (rare) cases in which the

hæmorrhage appears to be primary the blood is poured out from the thin-walled vessels of a gliomatous tumour. In all cases in which a spinal paralysis is developed with such rapidity as to suggest hæmorrhage, the patient and his friends should be closely questioned as to the presence or absence of previous symptoms suggestive of a commencing myelitis, on the one hand, or of a pre-existing chronic spinal lesion such as a glioma, on the other.

Prognosis.—You will readily understand that it is very difficult, indeed impossible, to lay down any general rules of prognosis which will apply to all cases of acute myelitis, for the disease depends, as we have seen, on many different causes and consists of many varieties, each of which presents great clinical differences in regard to their severity and course. But speaking generally, we may say that the prognosis as regards complete recovery is very unfavourable. Some cases (I refer to the more severe forms of indiscriminate myelitis) die during the acute stage of the disease. Death is usually due to paralysis of the respiratory muscles or extension of the inflammation to the medulla oblongata. Hence, the danger to life during the acute stage of the disease is much greater in cases of diffuse central and disseminated than in cases of local and transverse myelitis.

Other things being equal, a transverse myelitis in the dorsal region is more favourable than a transverse myelitis in the lumbar region; for a myelitis of the lumbar enlargement is more likely to be attended with severe paralysis of the bladder and the development of bed-sores. The acute (trophic) form of cystitis and the acute (trophic) bed-sore are very serious and dangerous complications. Further, a transverse myelitis of the lumbar enlargement is usually more favourable than a transverse myelitis of the cervical enlargement, for in the latter case the respiratory muscles are apt to be involved, and the inflammation is more likely to extend to the medulla oblongata. Fortunately, transverse myelitis in the cervical region (I exclude, of course, the secondary forms) is almost unknown.

In the great majority of cases of acute transverse myelitis, even when the inflammation is severe, the patient recovers from the acute stage of the disease, provided only that he is properly doctored and nursed. It is very important to remember that the

prognosis in acute transverse myelitis largely depends upon the circumstances and surroundings of the patient and upon the way in which he is treated and nursed.

When the acute stage is passed, the immediate risk to life is at an end. The degree of improvement and the extent of recovery which will probably take place have then to be considered. In trying to determine this point in each individual case, numerous circumstances have to be taken into account. The more important are:—The extent and severity of the inflammation, the part of the cord which is involved, the condition of the cord (whether healthy or not) prior to the onset of the myelitis, the cause of the myelitis (i.e. whether it is traumatic, idiopathic, syphilitic, due to compression, etc.). Some of these points, such as the extent and severity of the paralysis, are so obvious that they need not be further insisted upon.

The extent and severity of the damage to the grey matter—and we judge of this, you will remember, by observing the condition of the paralysed muscles as regards rapid atrophy, electrical excitability and reflex action, and the exact character of the paralysis of the bladder and rectum—is a most important point. In a case of transverse myelitis in which the grey matter of the lumbar enlargement is extensively involved, the prognosis as regards recovery from the paralysis is, other things being equal, much less favourable than when the dorsal region is involved; for nerve cells which are once degenerated and destroyed are, so far as we know, never restored; but there seems good reason to suppose that nerve tubes which are degenerated are not unfrequently restored.

When I come to describe compression myelitis and the spinal lesions which result from traumatic injury, I will refer in detail to some cases which have come under my own notice in which very great improvement, and indeed in some cases complete cure, has resulted from severe transverse myelitis—not merely, you will observe, from compression of the cord, but from undoubted myelitis associated with such compression. Acute (idiopathic) transverse myelitis when severe is, of course, a very different thing from a pressure myelitis which is generally subacute; a severe acute transverse myelitis produces, as we have seen, great destruction of tissue and is usually followed by

much cicatricial and sclerotic thickening which interferes with the restoration and regeneration of the nerve tubes. The same amount of restoration cannot be expected in cases of this kind as in cases of compression myelitis; but a certain amount of restoration does in many cases undoubtedly take place.

In syphilitic cases, the prognosis is perhaps somewhat more hopeful than in other forms of myelitis; but there are many exceptions to this general statement, for the syphilitic poison may lead to the production of myelitis and paraplegia in several different ways, and the prognosis is by no means the same in all cases. When the myelitis is due to the presence of a gumma on the surface of the cord, the prognosis is much more favourable (provided that active antisiphilitic treatment is employed at a sufficiently early period of the case) than in those cases in which the paraplegia is the result of syphilitic disease of the blood-vessels. Very remarkable recoveries are sometimes seen in syphilitic cases; the following case is one of the most striking which has come under my own notice:—

Case of acute myelitis; paraplegia; paralysis of the bladder; septic cystitis; rapid improvement and complete cure under iodide of potassium.—On February 6th, 1891, I saw a gentleman aged 42, who was suffering from acute myelitis. His bladder and lower limbs were completely paralysed; the urine was ammoniacal and loaded with blood, pus and phosphates.

Twelve years previously, he had contracted syphilis; the cicatrix of a large chancre was present on the body of the penis.

Five weeks before my visit, he had got very wet while curling on ice covered with water. During the three weeks succeeding the wetting, he suffered from pain in the back, shooting pains round the waist and a peculiar sensation of constriction and cold round the abdomen (girdle sensation); he then began to have difficulty in emptying the bladder (paralysis of the detrusor muscle); this was followed by weakness and numbness in the legs and sleeplessness.

Four days before my visit, the loss of power in the legs and the paralysis of the bladder had rapidly increased. On several occasions he had felt chilly, but there had been no fever.

I found the lower extremities completely paralysed, the muscles soft and flabby but not markedly atrophied, the knee-jerks and the plantar reflexes markedly diminished but not completely abolished. Sensibility to pain was completely abolished in the right leg and very markedly impaired in the

left; thermal sensibility was considerably impaired in both legs; the tactile sensibility in both lower extremities was only slightly affected. A broad band of hyperæsthesia extended round the abdomen and lower part of the chest; its vertical extent was about six inches, its lower border was situated about an inch above the umbilicus.

The bladder was completely paralysed and the urine very ammoniacal. The bowels were obstinately constipated.

There was no pain in the spine. A pink blush, suggestive of a commencing bed-sore, was present over the middle of the sacrum and adjacent parts of the buttocks. The vasomotor condition of the skin of the abdomen was obviously disturbed; scratching the skin with the point of a blunt tuning-fork produced a marked blush, and a blister had formed at a point at which a warm, but not a hot, poultice had been applied.

The pulse numbered 96 per minute; the temperature was normal; the tongue was markedly furred and indented with the teeth.

The case was clearly one of acute transverse myelitis; the exciting cause appeared to be exposure to cold and wet; the previous syphilis was thought to be in all probability an important factor in the case.

It was agreed to place the patient on a water-bed; to instruct the nurse to keep the back and adjacent parts scrupulously clean and dry; to wash out the bladder with boracic acid solution; and to give boracic acid (twenty grains three times daily), iodide of potassium (ten grains, subsequently increased to twenty grains, three times daily) and liquid extract of ergot (twenty minims three times daily) internally.

Subsequent Progress of the Case.—For three or four days after my visit, there was no improvement, in fact the reverse; the urine continued to dribble away constantly and remained ammoniacal and loaded with pus and blood; the patient passed into a semi-comatose condition and seemed about to die. Improvement then began to occur; the ammoniacal condition of the urine disappeared and the general condition improved; the sphincter of the bladder regained its tone, but retention lasted for some time.

At the end of four weeks, during the whole of which time the bladder was diligently washed out several times a day with boracic solution, the patient could move his toes. After this date (March 7th) the improvement was very rapid.

On *March 28th*, the patient was able to stand. On *April 6th*, he was able to walk. On *May 10th*, he walked a distance of about five miles over a hilly country without any bad result.

On *June 24th*, he came in to Edinburgh to see me. He was then looking quite strong and well. A few days previously, he had walked ten miles over the hills without any inconvenience or difficulty. No trace of paralysis could be detected in the lower extremities. The muscles were firm and well nourished; the patient could walk backwards with ease and could stand steadily and securely on either leg with the eyes closed. The knee-jerks were markedly exaggerated and there was a slight tendency to ankle-clonus. The bladder still continued at times to give him some inconvenience; there was still at times difficulty in expelling water. The patient was still taking the iodide and boracic acid (twelve grains of each three times daily).

The improvement has steadily continued, and for the past two years the patient has been absolutely well.

It is probable, I think, that in this instance the myelitis was due to a coarse syphilitic lesion—a gumma. It is difficult to explain the very remarkable and complete recovery on any other supposition.

It must, however, be remembered that syphilitic paraplegia is, in some cases, uninfluenced or very little benefited by anti-syphilitic treatment. The explanation is that in these cases the paralysis (the paraplegia) is an indirect result of the syphilitic lesion. If the paraplegia is due to destruction of tissue (softening) the prognosis is hardly if at all more favourable in syphilitic than in other cases; for iodide of potassium and mercury are unable to restore nerve tissue which is destroyed. I shall return to this point more in detail when I come to speak of cerebral syphilis and of the hemiplegic form of paralysis which is so frequently due to syphilitic disease of the cerebral vessels and the cerebral softening which results therefrom.

Let me repeat that, in cases of paraplegia due to transverse myelitis, the prognosis as regards the rapidity and extent of recovery depends to a considerable extent upon the position of the lesion.

In most cases of transverse myelitis involving the dorsal region, the recovery is sufficiently complete to enable the patient to walk, it may be with the help of sticks; and in some cases the recovery is much more considerable even than this. It is different when a severe transverse myelitis involves the lumbar enlargement. In cases of this kind, the prognosis as regards the

paralysis of the muscles of the lower extremities is very similar to the prognosis in cases of poliomyelitis anterior acuta involving the grey matter of the lumbar segments of the cord; but it is very important to remember that in cases of acute indiscriminate myelitis there are additional risks due to the paralysis of the bladder and rectum and to the formation of bed-sores. I have repeatedly told you, and I make no apology for insisting upon it again, that the paralysis of the bladder is one of the most important points which has to be considered both for the purposes of prognosis and treatment in this and indeed in many other affections of the spinal cord.

In many cases of acute myelitis, the paralysis of the bladder remains as a permanent condition; and in cases of chronic paraplegia the prognosis largely depends upon the degree and form of the bladder paralysis which is present. I need not say that in cases of chronic paraplegia the comfort of the patient largely depends upon the condition of the bladder. It not unfrequently happens that patients who have recovered sufficiently to go about and be able to attend perhaps to business, ultimately die of kidney and bladder complications, the result of paralysis of the bladder and septic cystitis. I remember one case of this kind which made a very profound impression on my mind. The patient, a gentleman, the manager of a large Insurance Company in the north of England, had a severe transverse myelitis; in the course of time he recovered sufficiently to walk about with the help of sticks and fulfil his important business engagements. He remained in this condition of spastic paraplegia for several years, the bladder, however, being permanently paralysed; it was never emptied except by the aid of a catheter; a permanent condition of septic cystitis was established; the bladder became encrusted with thick masses of phosphates; and the patient ultimately died from uræmia and kidney complications. If it had not been for the bladder paralysis, I believe that he would in all probability have been alive to-day. I have seen several other cases of the same kind. As the result of my experience, I am satisfied that once the dangers of the acute stage of the myelitis are past, the prognosis, as regards longevity, depends far more upon the condition of the bladder than upon anything else.

In cases, then, of myelitis in which the bladder paralysis,

requiring the use of the catheter is present, and still more when the sphincters are paralysed, the prognosis is bad. But this statement must not be taken too absolutely. I have met with cases in which complete recovery has taken place, in which paralysis of the bladder and ammoniacal urine had lasted for several months. In one remarkable case which I will detail to you when I come to speak of the lesions of the spinal cord which result from traumatic violence, a paralysis of the bladder, not, however, requiring the use of the catheter, persisted for ten years and was then recovered from. Cases such as these are exceptional; but their occasional occurrence should be remembered in forming a prognosis. The longer I live and the more experienced I become, the more I believe in the recuperative and compensatory powers of Nature.

We have seen that the paralysis which results from myelitis may depend partly upon destruction of nerve elements, and partly upon a temporary interruption or arrest of function in the nervous tissues, the result of shock, derangement of the circulation, and the pressure which the inflammatory œdema and exudation produce. Now, in cases of transverse myelitis involving the dorsal portion of the cord, it is impossible to say in the early stages of the case how much of the paralysis is due to destruction and how much is due to a merely temporary arrest of function. The extent and degree of the paralysis may afford no information; the only method of forming an opinion is to watch the course of events. Of course, in those cases in which the paralysis is slight, the prognosis is more favourable than in those cases in which the paraplegia is complete; but complete paraplegia is not necessarily indicative of destruction of tissue. In cases of transverse myelitis involving the lumbar enlargement, the electrical condition of the paralysed muscles is of considerable importance. It is unnecessary to say that in those cases in which the reaction of degeneration is developed the prognosis is much less favourable than in those cases in which the electrical reactions remain normal or undergo a simple diminution. We have considered this point so fully in connection with poliomyelitis anterior acuta that I need not enter into details.

Further, we have seen that in most cases of myelitis sensation is less severely affected than motion, and that the sensory

paralysis is more quickly recovered from than the motor paralysis. In those cases in which the anæsthesia is very profound and in which the sensory paralysis remains, after the acute stage is past and a reasonable time has been allowed for the development of reparative changes, the prognosis is unfavourable; *vice versâ*, in those cases in which the anæsthesia is slight and temporary, the prognosis is more favourable. The same statement applies to the motor paralysis. If there is no return of motor power, say in the toes, at the end of three months, the prognosis is very bad; and the longer the persistence of complete paralysis the worse, of course, the prognosis.

LECTURE XXVIII

ACUTE MYELITIS (*Continued*)

TO-DAY, Gentlemen, I wish in the first place to direct your attention to the treatment of acute myelitis.

Treatment.—The objects of treatment are:—To cut short the inflammation and to prevent its development and extension; to tide the patient through the acute stage of the disease; to promote recovery and the restoration of the damaged nerve elements, once the acute stage is past; and as far as possible to avoid, during the subsequent stage of chronic paraplegia and spastic paralysis, everything which is likely to re-excite the inflammation or to produce complications.

It is very doubtful whether we are able in any material degree to influence (arrest) the inflammation in its early stages. Still, if the case is seen sufficiently early, an attempt should be made to restrain the vascular changes which precede and attend the stage of inflammatory exudation.

With this object, a brisk purge should be administered at the beginning of the attack, and leeches, cups (dry or moist), an icebag or hot fomentations¹ should be applied over the affected part of the spine. A febrifuge mixture may also be prescribed. Dry cupping is probably the most effective local application.

Attention to position is of some importance. It is advisable if possible to prevent the patient lying continuously on his back; if it can be managed, he should be encouraged, from time to time, to lie on his side or on his face, so as to prevent the congestion of the cord which is apt to be produced when he lies continuously on his back.

¹ I need not again discuss the question whether cold or heat is the more effective in restraining the vascular changes which attend a commencing inflammation. I have already referred to this point in connection with the treatment of poliomyelitis anterior acuta. (See page 95.)

It is very essential in all cases of myelitis, and particularly in severe cases where there is a great risk of bed-sores, to place the patient from the first on a water-bed. Some patients find it difficult to lie on the side on a water-bed; in such cases we must be content to allow the patient to lie on his back. I cannot too strongly insist on the importance of placing the patient on a water-bed, for one of the most important points in the treatment of acute myelitis is to prevent the development of bed-sores. The back, hips, knees and ankles—the points where bed-sores are most likely to form—should be inspected daily, not only by the nurse, but also by the doctor.

Ergot, belladonna, tartar emetic, quinine, salicylate of soda, iodide of potassium, mercury and many other remedies have been recommended during the acute stage. Quinine, iodide of potassium and mercury are probably the most useful drugs.

The diet must be light and nutritious, and the sick-room should be well ventilated and kept at an equable temperature.

The most scrupulous attention should be given to cleanliness. And this leads me to say that one of the most important points in the treatment of acute myelitis is to attend to the condition of the bladder and rectum. We have seen that in some cases the sphincters are paralysed, and that in others there is unconscious reflex urination. In consequence of the anæsthesia, the patient may be quite unconscious that the urine is escaping. If the patient is allowed to lie wet and dirty, it is very difficult to prevent the formation of bed-sores. One of the most important duties of the nurse is to see that the patient is dry and clean. Anything which is likely to produce peripheral irritation should, so far as possible, be avoided. If it is thought necessary to place hot bottles in the bed, care should be taken that they are not too hot and that they are not allowed to come in direct contact with the paralysed parts. In consequence of the lessened resisting power of the skin, a degree of warmth which would only be pleasant to a healthy individual may cause inflammation and sloughing. Further, in consequence of the anæsthesia, the patient is unable to feel whether the bottle is too hot or not. The nurse and the doctor have in such cases to feel for him.

If it is necessary to use the catheter for the relief of the retention, the greatest care should be taken to see that it is clean. We have seen that in some cases, notwithstanding the

most scrupulous aseptic precautions, the urine becomes ammoniacal and a severe form of cystitis is developed, apparently as the result of a trophic lesion in the wall of the bladder.

When cystitis is developed, it is of great importance to endeavour to allay it as speedily as possible. With this object, the bladder should be frequently washed out with a weak, slightly warm solution of boracic acid; and boracic acid in twenty-grain doses should be given internally. To allay the vesical irritation, morphia suppositories may be freely employed.

After the more acute stage has subsided, the same treatment which has been recommended for poliomyelitis anterior acuta should be employed. During the early stages of the case, rest is essential, for it is important to avoid irritation of the damaged nerve elements. Our object is to promote absorption of the inflammatory products and to aid the reparative process. Powerful attempts to produce voluntary movements and reflex stimulation (the application of the faradic battery, etc.) are apt to do harm rather than good. During this stage, iodide of potassium or iodide of potassium and mercury, the syrup of the iodide of iron, quinine and general tonics may be prescribed. At this stage, electricity and strychnine are to be avoided.

In the course of a month or six weeks, depending upon the severity of the case and the condition of the paralysed muscles, judicious massage and electricity, and above all, systematic, voluntary efforts, may be employed.

In those cases in which there is much rigidity and tension of the muscles and great exaggeration of the deep reflexes, the application of the faradic current to the paralysed limbs and the administration of strychnine are apt to do harm rather than good. When these conditions (rigidity and tension of the muscles, and exaggeration of the deep reflexes) are associated with a very considerable degree of atrophy—and in some cases the two conditions are combined—a weak faradic current and small doses of strychnine or *nux vomica* may be employed in combination with hydrobromic acid. A double effect is produced in this way; the hyperexcitability, so to speak, of the reflex centres is calmed by the hydrobromic acid, and the enfeebled nerve cells (they are in a condition of irritable weakness) are toned up, as it were, by the strychnine or *nux vomica*.

When the muscles are flaccid, strychnine, the faradic current and massage are the most appropriate remedies.

In both classes of cases (whether the paralysed limbs are spastic or flaccid), arsenic and general tonics, such as quinine, iron, the compound syrup of the phosphates, cod-liver oil, etc., may be prescribed. Good feeding and plenty of fresh air are also, of course, desirable after the acute stage is passed.

It is doubtful if counter-irritation applied to the spine over the seat of the lesion has any beneficial effect upon the progress of the disease; possibly in some cases in which the spastic symptoms are very prominent the actual cautery is useful.

I need not repeat what I have already stated (see p. 100) with regard to the effect of the constant current applied to the spine further than to say that a weak constant current, if judiciously employed, can, in my opinion, do no harm; but I doubt if it is likely to do much good.

If the bladder still remains paralysed after the chronic stage of the disease is reached, it is of the utmost importance to impress upon the patient the paramount importance of continued care in the use of the catheter, and the avoidance of everything which is likely to establish cystitis and to induce kidney complications. When the urine continues ammoniacal, boracic acid and quinine should be administered, the bladder should be diligently washed out with a weak solution of boracic acid and (unless the other symptoms contra-indicate its use) strychnine should be given internally. In cases of this kind, the bladder may be faradised as soon as the cystitis is allayed, one pole being placed in the bladder (which has previously been filled with water) and the other over the pubes or perineum.

In severe cases of myelitis, recovery is usually very slow; it may be many months before the motor power is sufficiently restored to enable the patient to stand or to walk even with the help of sticks. During this stage of the disease, it is of the greatest importance, provided that some attempt at repair, however slight, is going on, to sustain the patient's mental tone and to encourage him to hope that in the course of time he will be able to stand and walk. At this stage of the disease, no tonic is so powerful as a mental tonic—the hopeful frame of mind which a favourable opinion produces; and no means of promoting repair and restoring the motor power is so effective

as frequently repeated and systematic voluntary efforts to move the paralysed parts. So long as some improvement is going on, the patient should be encouraged, day by day, to hope for more. Whenever recovery has advanced sufficiently far to enable the patient to stand, even with support, he should be encouraged to try and walk. I have more than once told you that during the early stage of the case, when the acute changes are still in active progress, attempts at voluntary movement should be avoided. But, once the acute stage has subsided, say at the end of six or eight weeks, the patient should be encouraged to make regular systematic movements of the paralysed muscles, and when a certain amount of motor power has been regained he should be encouraged to attempt to stand and to try to walk. An enormous step is gained when the patient finds that he is able to stand, and still more when, with support and help, he is able to walk a few steps across the floor. He then feels himself in an altogether different position; he looks with renewed hope to the future; he realises that he *will* ultimately be able to walk. Although, in many cases of transverse myelitis, the patient recovers sufficiently to be able to walk, recovery is very rarely complete when the myelitis has been severe. In cases of this kind, a condition of spastic paraplegia usually remains.

The treatment of the spastic paraplegia which results from acute myelitis is practically the same as that of primary spastic paralysis, which we have already considered. The pathological process is of course somewhat different in the two conditions. In primary spastic paraplegia, sclerosis of the crossed pyramidal tract is gradually and slowly developed in a healthy cord; whereas in secondary spastic paraplegia, the sclerosis is a secondary degeneration developed as the result of an acute lesion in a cord which is diseased. But notwithstanding this difference, the treatment is practically the same in the two conditions. There is, however, one important difference. In primary spastic paraplegia, the bladder is rarely affected in any marked degree; but in secondary spastic paraplegia following acute myelitis, the bladder is often paralysed. This, as I have already more than once pointed out, is a fact of the greatest importance. The condition of the bladder always requires the most scrupulous care and treatment.

The above remarks chiefly apply to those cases of acute

transverse myelitis in which the lesion is situated in the lower dorsal or lumbar portions of the cord. When the upper dorsal or cervical segments are affected (but this is so rare that some authorities seem to doubt its occurrence) the condition of the respiratory muscles requires great attention. Paralysis of the respiratory muscles is a source of grave danger; and in all cases of myelitis in which the respiratory muscles are affected, it is important, both during the acute and also during the subsequent chronic state of the disease, to avoid everything which is likely to throw a strain on the respiratory apparatus. When the respiratory muscles are paralysed, a trivial bronchitis may prove fatal. In cases of this kind, a common cold, because of its liability to be followed by bronchial catarrh, should be carefully treated.

In the diffuse and central forms of myelitis, our therapeutic efforts are rarely attended with any measure of success. In these cases, the inflammation usually advances with great rapidity and a fatal termination is often reached within a few days from the onset of the attack.

In the disseminated form of myelitis the prognosis is also very bad and treatment very ineffectual. In some of these cases, the inflammation is, as we have seen, a result of syphilis; and in cases of disseminated myelitis, whether there is a definite history of syphilis or not, it is always, I think, advisable to try the effect of iodide of potassium and mercury. The same statement applies to many other forms of subacute and chronic myelitis. But you must remember that even in those cases of subacute and chronic myelitis in which there is a distinct history of syphilis, antisyphilitic remedies often fail to give relief. I have known very grave complications—the extension of the inflammation to the medulla oblongata, for example—actually develop while the patient was under a course of antisyphilitic treatment. The risk of extension of the inflammatory process to the medulla oblongata is, in my experience, much greater in the central and disseminated than in the other forms of myelitis; but the possibility of its occurrence must be kept in mind in all forms of the disease.

Some years ago I saw, with Dr. Playfair, a lady who was suffering from what appeared to be an early stage of primary spastic paraplegia. She had repeated attacks of recurrent

herpes of the vulva, a condition which is often a result, though not usually the direct result, of syphilis, but which is not itself directly influenced by antisyphilitic remedies. In her case, iodide of potassium and mercury did not produce any beneficial effect upon the cord lesion, although there was good reason to believe that it was specific. In her case, bulbar symptoms suddenly developed and she died in the course of a few days from what appeared to be an acute inflammation of the medulla oblongata.

CHRONIC MYELITIS

THE next subject to which I propose to direct your attention is chronic myelitis. In cases included under this term, the lesion is indiscriminate, and the course slow and usually progressive. Unless as the result of bladder or other complications, there is no fever.

Etiology.—In some cases, chronic myelitis is secondary to an attack of acute myelitis; and the same causes which give rise to acute myelitis seem to produce the chronic form of the disease. Alcoholic excesses, syphilis, exposure to cold and traumatic injury seem to be the most frequent causes of the condition. The influence of cold and traumatic injury has probably been overrated. In many cases the symptoms develop without any apparent cause; such cases are sometimes termed idiopathic.

Compression of the spinal cord is apt to produce a distinct form of myelitis to which I will afterwards refer in detail. (See page 570.)

Morbid Anatomy.—The extent and distribution of the lesion are subject to great variations in different cases. As in the acute form, several distinct varieties may be described; the more important are:—(a) The local or focal; (b) the transverse; and (c) the disseminated forms.

On macroscopic examination, the affected parts of the cord are usually firmer than normal, the increased density being due to sclerosis. In advanced cases the nerve elements may be entirely replaced by connective tissue.

On microscopic examination, the same changes which have been described as characteristic of the third stage of acute myelitis are present. There is, in short, no sharp line of demarcation between the terminal period of an acute attack and the chronic form of myelitis. In the affected parts of the cord, the nerve tube may, in places, have completely disappeared.

The connective tissue trabeculae are thickened; the lymphatic sheaths are often dilated; the Dieters' cells are larger and more numerous than in health. In those parts of the cord in which the lesion is less advanced, enlarged axis-cylinders may be seen. The appearances, of course, vary with the extent and severity of the lesion, and the duration of the case. In some cases, the whole or the greater part of the transverse section of the cord appears to be disorganised, its proper structures having entirely disappeared, and being replaced by connective tissue; the blood-vessels in the affected area are usually dilated and their walls thickened.

Clinical History.—In cases which are chronic from the first, the onset is, as a rule, extremely slow and gradual. The first symptom of which the patient complains is usually motor weakness, sometimes a feeling of numbness, or 'pins and needles.'

The clinical picture presented by fully developed cases is subject to great variations. In the majority of cases, the motor symptoms are much more evident than the sensory; in fact in some cases there are no sensory derangements. The exact distribution of the motor weakness and paralysis, of course, depends upon the position and extent of the lesion. Both lower extremities are usually affected, but the loss of power is often much more marked in one than in the other. The muscles on the front of the leg and the peroneal muscles are in many cases severely affected. The deep reflexes are usually exaggerated. In many cases, there is both ankle-clonus and exaggeration of the knee-jerks. In exceptional cases in which there is no ankle-clonus, a knee-clonus can be elicited sometimes. In other cases, some of the affected muscles, especially the peroneal muscles and the muscles on the front of the leg are atrophied, and their electrical reactions conspicuously altered. In some cases, the patient complains of some loss of power in the upper extremities, and of numbness in the fingers and hands; but it is comparatively rare to find any well-marked paralysis or muscular atrophy in the upper limbs. In some cases there is little muscular wasting, the condition being practically one of spastic paraplegia. In fact, in many cases of primary spastic paralysis the lesion is probably a chronic myelitis.

Constipation is usually a prominent symptom.

There is often some derangement of the vesical reflex—inability to completely empty the bladder, dribbling of the last portions of the urine, precipitate urination, or even more marked paralysis.

Loss of sexual power is common in men.

The subjective disturbances of sensation are usually much more marked than the objective. In rare cases, there is extreme inco-ordination and loss of the muscular sense—an acute form of locomotor ataxia to which the term ataxic myelitis has been applied.

The general health is, as a rule, good, and the condition of the internal organs is usually quite healthy.

The course of the disease is very variable. In many cases, the paralysis slowly and gradually increases and the difficulty of walking becomes more marked, until ultimately the patient is confined to bed, unable to walk, or unable to move the lower extremities. In the terminal stages, the muscles may remain rigid; in others, the muscles below the knee are markedly atrophied, but the knee-jerks remain exaggerated.

Bladder complications (paralysis of the sphincter and cystitis) may be developed in the later stages of the disease; bed-sores not infrequently form in those cases in which the patient is not carefully nursed.

In some cases, especially those due to traumatic injury and to compression, the membranes are inflamed as well as the spinal cord itself. In these cases, symptoms characteristic of meningitis are present.

Diagnosis.—The diagnosis of chronic myelitis is in many cases synonymous with the diagnosis of chronic paraplegia. The physician has, in the first place, to determine whether the paralysis is due to an organic or functional lesion (see page 142); and in the second place, if an organic lesion (myelitis) seems to be present, to endeavour to come to a conclusion as to the primary cause of the cord lesion. In all cases of chronic myelitis, the possibility of the case being due to compression must be kept in view. I shall return to this point in more detail when I come to describe compression myelitis.

The exact position of the lesion in the cord, its height and

distribution in the transverse section are of course determined by observing the exact distribution of the motor and sensory derangements, and of the trophic and reflex alterations which happen to be present.

Prognosis.—The course of the disease is usually very protracted, but in most cases the lesion slowly progresses and the difficulty in walking and other symptoms gradually and slowly increase. Occasionally the morbid process is arrested and the motor paralysis is to some extent recovered from. Recovery is rare, except in syphilitic cases, and even in the syphilitic cases a *complete* cure rarely results. In some cases, temporary improvements and repeated relapses occur. It is unnecessary to say that in cases of this description the differential diagnosis from functional (hysterical) paralysis may be exceedingly difficult.

Treatment.—The same measures of treatment which have been recommended for the paraplegia which follows acute myelitis should be adopted. The most useful remedies are iodide of potassium and mercury; nitrate of silver, arsenic, quinine, strychnine, hydrobromic acid and phosphorus. Iodide of potassium and mercury are, of course, especially valuable in syphilitic cases. I need hardly say that strychnine or nux vomica are apt to do more harm than good in those cases in which the spastic symptoms are very prominent. In many of the cases in which some of the muscles, such for example as those on the front of the leg, are paralysed and atrophied, and in which the deep reflexes are exaggerated, a combination of hydrobromic acid with small doses of strychnine or nux vomica is valuable. In cases of this kind, massage and electricity are also beneficial. The constant current may be applied to the spinal cord. Erb speaks very favourably of a properly conducted water-cure and of electricity.

Counter-irritation to the spine, especially the application of the actual cautery, is undoubtedly useful in some cases.

ACUTE ASCENDING PARALYSIS: LANDRY'S PARALYSIS

UNDER the name 'paralysis ascendens acuta' Landry directed attention to a rare affection, the chief characteristic of which is motor paralysis which commences in the muscles of the lower extremities, and which gradually and usually with great rapidity extends upwards, involving the muscles of the trunk and upper extremities, and it may be finally the muscles supplied by motor nerves rising from the medulla oblongata. The paralysis is a flaccid paralysis; the reflexes are usually abolished; even when the patient lives long enough, rapid atrophy and the reaction of degeneration are not developed.

Etiology.—The exact conditions which predispose to, and which excite, this rare disease are unknown. The affection seems to be more common in men than in women. It usually occurs between the ages of twenty and forty. Children seem to be very rarely affected. In some of the cases which have been reported, the patients had been exposed to cold and wet; in others there was a history of syphilis or of alcoholic excess; in others again, the disease followed an acute febrile affection, such as typhoid fever or pneumonia, a septic disease, or traumatic injury.

Morbid Anatomy.—In most of the cases which have been carefully examined post mortem the spinal cord, peripheral nerves, muscles, in short the whole of the neuro-muscular apparatus, seemed to be healthy. In a few cases, changes which were thought to be indicative of commencing inflammation were present in the grey matter of the spinal cord or in the peripheral nerves. But in the majority of cases, as I have just told you, the post-mortem results were entirely negative.

Pathology.—The absence of any obvious lesions in the neuro-motor apparatus, and the fact that in some cases the

spleen presents changes similar to those which are met with in the infectious fevers have led Landry and some other observers to suppose that the disease is due to the introduction into the system of some toxic material. The fact that the disease seems in several cases to have been preceded by an acute febrile affection or a septic inflammation, lends some support to this view.

Schulz and Schultze have suggested that acute ascending paralysis may perhaps represent the slightest form of a general and spinal bulbar inflammation in which the pyramidal fibres are specially affected.

Gowers thinks that the lesion is probably situated at the terminal fibres of the pyramidal tract—the fine fibrils in which the fibres of the pyramidal tract terminate in the grey matter of the spinal cord and bulb. A lesion in this situation might, he points out, be expected to produce motor paralysis and loss of reflex action without causing muscular atrophy and the reaction of degeneration. The fact that a chronic lesion in this position produces a spastic rather than a flaccid paralysis is not perhaps of much importance in opposition to this view; for this disease is essentially acute.

Clinical History.—In some cases, the actual onset of the paralysis is preceded by premonitory symptoms, such as slight febrile disturbance, numbness, weakness, heaviness in the limbs. In others, the loss of power is developed rapidly.

The characteristic symptom is muscular weakness which quickly becomes complete paralysis.

As a rule, the muscles of the toes and feet are first involved. Those of the leg and thigh are next affected, then those of the trunk, hands and arms. Finally the diaphragm and muscles supplied from the medulla oblongata may be involved; difficulty in breathing and in swallowing may consequently be developed in the later stages of the case; the patient not unfrequently dies asphyxiated.

Although the paralysed muscles are flaccid, they do not undergo any marked and rapid atrophy; and, even if the patient should live long enough to permit of its development, they do not present the reaction of degeneration.

In the early stages of the case, the reflexes may be unaltered;

as the disease progresses, they usually become diminished and finally abolished.

In the great majority of cases, sensory disturbances are either entirely absent or only very slightly marked.

The bladder and rectum are seldom affected. Trophic alterations in the skin do not occur. It is only in rare and exceptional cases that there are any cerebral symptoms.

In many cases there is no fever and no constitutional disturbance. In others, the temperature is elevated and the usual constitutional disturbances which attend an acute febrile process (quick pulse, thirst, emaciation, etc.) are developed.

Profuse sweating is a common symptom; the spleen is usually enlarged; swelling of the lymphatic glands sometimes occurs.

Duration, Course and Termination.—The duration varies considerably in different cases, the average being from eight to twelve days. The disease not unfrequently proves fatal in from three to four days, indeed in some cases within forty-eight hours, from the commencement of the attack. In other cases, the fatal termination is not reached for two or three weeks. A fatal result is the rule; it would appear that in rare and exceptional cases recovery occasionally takes place.

Diagnosis.—The more important facts from a diagnostic point of view are:—The rapid development and flaccid character of the paralysis, the abolition of the reflexes, the absence of marked atrophy and of the reaction of degeneration in the paralysed muscles, the absence of any affection of the bladder or rectum and of marked disturbances of sensation.

Further, the facts that the paralysis usually commences in the lower limbs, that it rapidly extends upwards, and that in many cases it finally involves the medulla oblongata are points of great importance.

The conditions most likely to be confounded with Landry's paralysis are:—Acute myelitis, poliomyelitis anterior acuta, peripheral neuritis and hysterical paraplegia.

The differential diagnosis of Landry's paralysis and of the common forms of acute myelitis.—The ordinary form of acute myelitis is very rarely limited to the motor side of the nerve apparatus;

consequently the presence or absence of sensory derangements is an important point of distinction. In Landry's paralysis there are no marked disturbances of sensation ; the bladder and rectum are unaffected, and there is no tendency to the formation of bed-sores ; while in the ordinary form of acute myelitis, sensory derangements, paralysis of the bladder and rectum and bed-sores are usually conspicuous features.

Further, the condition of the muscles and of the reflexes in Landry's paralysis is essentially different from that which is observed in cases of transverse myelitis affecting either the lumbar or the dorsal region of the spinal cord.

Cases of acute central myelitis which pursue a rapid and ascending course bear a much closer resemblance to Landry's paralysis. Indeed, in some cases the diagnosis may be impossible. The absence of sensory disturbances (especially of any derangement of the sensibility to pain, heat and cold) of muscular atrophy and of bed-sores and of any derangement of the functions of the bladder and rectum are the most important points.

The differential diagnosis of Landry's paralysis and of poliomyelitis anterior acuta.—This presents no difficulty. Landry's paralysis very rarely occurs in children ; while poliomyelitis anterior acuta very rarely occurs in adults. In Landry's paralysis, the course of the disease is progressive ; while in poliomyelitis anterior acuta, the height of the paralysis is at once reached. In Landry's paralysis (provided that the patient survives long enough for these conditions to be developed), rapid atrophy and the reaction of degeneration are never observed ; but in poliomyelitis anterior acuta, rapid atrophy and the reaction of degeneration are conspicuous and characteristic features. Further, Landry's paralysis is usually fatal ; while poliomyelitis anterior acuta is very rarely (if ever) directly fatal.

The differential diagnosis of Landry's paralysis and of peripheral neuritis.—This is sometimes more difficult, but in the majority of cases a correct decision can be arrived at. In Landry's paralysis, the onset of the paralysis and the course of the disease are usually more rapid than in peripheral neuritis. In Landry's paralysis, there is an absence of marked atrophy, of the reaction of degeneration, of tenderness over the nerve trunks, and of definite symptoms indicative of sensory derangement, all of which conditions are usually well marked in cases of peripheral

neuritis. Further, the mode of development and spread of the paralysis in the two cases is different. In the majority of cases of Landry's paralysis, the paralysis begins in the toes and gradually extends upwards to the muscles of the legs, thighs, trunk and upper extremities. In cases of peripheral neuritis in which both the lower and upper extremities are involved, the paralysis usually affects the peripheral parts of the upper and lower extremities (hands and feet) more or less simultaneously. As I have already stated, some observers believe that the fundamental lesion in Landry's paralysis is a peripheral neuritis. I see no reason to accept this view. At the same time, it must be admitted that the affection of the spinal cord which is in all probability the pathological substratum of Landry's paralysis (a toxic disturbance of the motor tracts and anterior cornua) is not unlikely to be associated with, and perhaps to be the cause of, inflammatory or degenerative changes in the peripheral nerves.

In this connection I may mention that Déjerine found alterations in the anterior motor nerve roots which he regarded as secondary to changes of a trophic kind in the multipolar nerve cells of the anterior cornua of the spinal cord—alterations so delicate that they cannot be discovered by our present means of histological observation.

In many of its features Landry's paralysis closely resembles the subacute inflammation of the anterior cornual region of the spinal cord which was described by Duchenne under the term '*paralysie générale spinale antérieure subaiguë*.' The chief points of distinction between the two conditions are:—the mode of onset and development of the paralysis, the condition of the paralysed muscles, and the clinical course. (See page 111.)

Prognosis.—This is very unfavourable. The great majority of cases terminate fatally. Cases which pursue a rapidly ascending course, with early implication of the respiratory muscles and the development of bulbar symptoms, are almost certain to prove fatal. In every case of acute ascending paralysis, whether due to Landry's paralysis or to any other cause, such as central myelitis, the prognosis must always be very guarded, even although the symptoms at the commencement appear to be comparatively slight.

Treatment.—The same treatment which has been recommended for acute myelitis and poliomyelitis anterior acuta should be adopted. Further, it should be remembered that the condition is in all probability the result of some toxic cause. Quinine, tincture of the perchloride of iron, boracic acid and other remedies which are useful in septic and toxæmic conditions are consequently indicated.

LECTURE XXIX

MULTIPLE CEREBRO-SPINAL SCLEROSIS¹

THE next subject, Gentlemen, to which I wish to direct your attention is the interesting disease to which the terms *Multiple Sclerosis*, *Multiple Cerebro-spinal Sclerosis*, *Disseminated Sclerosis*, *Insular Sclerosis*, *Sclérose en plaques disséminées*, and other synonyms have been applied.

Let me first refer to the morbid anatomy.

Morbid anatomy.—Multiple cerebro-spinal sclerosis is a chronic affection of the nervous system in which patches or islets of sclerosis are developed here and there in the nerve centres (brain, pons Varolii, medulla oblongata, cerebellum and spinal cord). In some cases, the peripheral nerves, more especially the cerebral nerves, and spinal nerve-roots are also affected.

The patches are for the most part situated in the white matter, although the grey substance, both in the spinal cord and brain, is occasionally involved. The lesion seems to have a special tendency to invade those parts of the nerve centres which consist of medullated nerve fibres. As we shall presently see, one of the most striking facts in the morbid histology of the disease is the disappearance of the medullary sheaths (white substance of Schwann) of the nerve tubes which are situated in the sclerotic areas.

In consequence of the fact that the white substance is the usual seat of the lesions, the sclerotic patches are often seen on the surface of the spinal cord, medulla oblongata, pons Varolii

¹ Although multiple cerebro-spinal sclerosis is a disease of the brain and spinal cord, it is convenient and advisable to consider it in connection with the diseases of the spinal cord. In some cases, the lesions are confined to the spinal cord, at all events during the earlier stages of the case. Cases of this kind cannot be intelligently studied and understood until the common (cerebro-spinal) form of the disease has been considered.

and crura cerebri, the surface of which is composed of white matter, but very rarely upon the exterior of the brain and cerebellum, the surface of which is composed of grey matter. But in the spinal cord, pons Varolii and medulla oblongata, the sclerotic patches are often best displayed, or only seen, after making a series of transverse sections.

The lesion is essentially an indiscriminate one. In the great majority of cases, the patches appear to be scattered at random, as it were, throughout the nervous tissue—especially the white matter; but occasionally symmetrically-placed patches are seen, more especially in the spinal cord. Possibly a symmetrical distribution of this description is merely accidental.

As I have just stated, sclerotic patches are often situated in the crura cerebri. The cerebellum is in some cases also involved—usually the white matter. In the brain, the patches are chiefly seen in the white matter of the centrum ovale, the corpus callosum, the crura cerebri, the walls of the ventricles and the septum lucidum; but the basal ganglia are not unfrequently involved. The grey matter of the convolutions is much less frequently affected.

Naked-eye appearances.—On naked-eye examination, the patches usually have a sharply defined outline; they stand out distinctly from the white matter by which they are surrounded; in the grey matter they are less distinctly seen.

The islets or patches of sclerosis vary from the size of a pin's head to that of a walnut; their usual size, as seen by the naked eye, is from a small pea to a hazel nut; the largest patches which I have seen have been situated in the centrum ovale of the cerebral hemispheres. In the spinal cord the individual patches rarely exceed the size of a hazel nut, and are not often so large as this. Adjacent patches sometimes run into one another, but this is not common. The patches usually have a grey or yellowish-grey colour; they are more or less translucent and glistening; after exposure to air they become somewhat pink or salmon-coloured. The sclerotic patches are firmer and tougher than the normal nerve tissue in which they are situated. On section, the surface of the sclerotic tissue sometimes occupies the same level as that of the normal nerve tissue by which it is surrounded; but in other cases it projects above or below that level. The different appearances as regards

colour, consistency and the level of the section seem to depend chiefly upon the age of the lesions.

Microscopical appearances.—On microscopical examination, it is found that some of the patches are sharply defined, while others gradually merge off into the surrounding nerve tissues. These differences are perhaps chiefly due to different methods of preparation. An essential feature of the lesion consists in the disappearance of the medullary sheaths of the nerve tubes. In sections prepared by Weigert's method (which stains the medullary sheath), the edges of the patches are usually very abrupt and sharply defined; but in carmine preparations (in which the neuroglial tissues are deeply stained) it is seen that the connective tissue overgrowth gradually merges into the surrounding healthy tissue.

Under a high power, the sclerotic patches are found to consist of an extremely delicate connective tissue in the midst of which naked axis-cylinders are usually present. The fibrillated appearance of the connective tissue is best seen in longitudinal sections, for the connective tissue bundles run parallel with the nerve tubes. The lesion, in fact, essentially consists of an increase of the neuroglial tissue—it is a neuroglial sclerosis. The Deiters' cells and connective tissue corpuscles are enlarged; the walls of the blood-vessels in the affected areas may be studded with nuclei, thickened and sclerosed; the vessels themselves are sometimes narrowed, sometimes, but less frequently, dilated. In some cases, compound granule corpuscles are present in great numbers, and crystals, which Frommann thinks are composed of margarine, are occasionally situated in the sclerotic areas. In those cases in which the compound granule corpuscles are abundant, fat globules and compound granule cells are often seen in the lymphatic spaces and adhering to the walls of the blood-vessels. These fatty granules are most abundant at the margins of the patches and in newly-formed patches; they undoubtedly represent the débris of the broken-down and degenerated medullary sheaths. Amyloid bodies are sometimes also present.

The condition of the nerve tubes varies in different situations and seems to depend upon the age and density of the sclerotic tissue which surrounds them.

At the edges of the sclerotic patches and in recently formed

patches, the chief alteration is an increase of the neuroglial tissue. In those parts at which the sclerosis is least marked, some of the nerve tubes may be perfectly healthy; in others the white substance of Schwann may have in great part disappeared and may be merely represented by a narrow band surrounding the axis-cylinder; the axis-cylinders themselves may be normal in size or hypertrophied. In those situations in which the sclerosis is more marked (i.e. nearer the centres of the sclerotic patches), the white substance of Schwann surrounding the axis-cylinders may have completely disappeared; some of the axis-cylinders, which then lie naked, as it were, in the midst of the hypertrophied neuroglia, may be enormously enlarged; others may be of normal size; while others may be atrophied. In the centres and densest portions of the patches, some of the axis-cylinders, like the white substance of Schwann, may have completely disappeared; but even in these situations many naked axis-cylinders are usually still present, while others may be atrophied. When the grey substance is involved, the nerve cells which are surrounded and encroached upon by the sclerotic overgrowth degenerate and atrophy.

In short, as the sclerosis advances, the nerve elements (fibres and cells) become more and more seriously affected, until finally some of the axis-cylinders and nerve cells may be destroyed.

From this description, you will see that the lesion essentially consists in:—(1) An increase or sclerosis of the neuroglial tissue with thickening and dilatation of the blood-vessels; and (2) the production of secondary changes in the nerve elements, viz., destruction and absorption of the white substance of Schwann, enlargement of the axis-cylinders which are laid bare and deprived of their protective and insulating sheath, and finally, it may be, atrophy and disappearance of some of the axis-cylinders and nerve cells.

In figs. 153 to 158, the cord changes which were present in a typical case of multiple cerebro-spinal sclerosis which came under my observation several years ago are represented. In that case, the sclerotic patches were very numerous; they were not confined to the spinal cord, but were scattered in great profusion through the medulla oblongata, pons Varolii, cerebellum, crura cerebri and cerebral hemispheres.

It is important to note that in many cases of cerebro-spinal

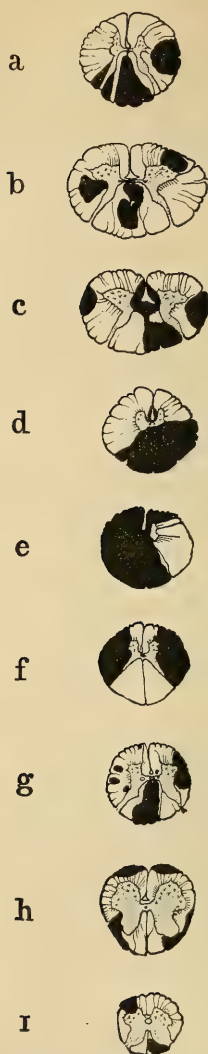


FIG. 153.—*Transverse sections through the spinal cord, from a case of cerebro-spinal sclerosis, showing the position and extent of the sclerotic (deeply stained) patches at different heights. About one and a half times the natural size.*

a, upper cervical region ; b, cervical enlargement ; c, cervical enlargement ; d, upper dorsal region ; e, mid-dorsal region ; f, mid-dorsal region ; g, upper lumbar region ; h, lower part of lumbar enlargement ; i, sacral region.

Sections b, d and h are shown more highly magnified in succeeding figures.

sclerosis, notwithstanding the presence of numerous sclerotic patches in the spinal cord, there is little or no secondary degeneration. The absence of secondary degeneration (ascending and descending) seems to be due to the persistence of the axis-cylinders. Those nerve fibres which are the upward or downward prolongation of axis-cylinders which have been completely destroyed will necessarily become degenerated.



FIG. 154.—*Transverse section through the cervical enlargement in a case of cerebro-spinal sclerosis. Stained with carmine, mounted in dammar, and magnified about 8 diameters.*

a, patch of sclerosis in the left lateral column; *b*, in the posterior column; and *c*, at the junction of the right lateral and anterior columns.

Etiology.—Multiple cerebro-spinal sclerosis is most frequently met with in young adults; it usually commences between the ages of twenty and thirty; it is very rare in childhood, practically unknown in infancy, seldom developed after the age of forty, and almost never met with in old age.

Most authorities agree in thinking that the disease is equally common in the two sexes; though Charcot was of opinion that females are more frequently affected than males. In the cases which have come under my own notice, the two sexes were almost equally affected.



FIG. 155.—Transverse section through the mid-dorsal region in a case of cerebro-spinal sclerosis. Stained with osmic acid, mounted in Farrant's solution, and magnified about 8 diameters.

The whole of the left half of the section (*b*), including the grey matter, is affected by the lesion. The posterior and anterior columns (*a*), on the right side, are also involved.

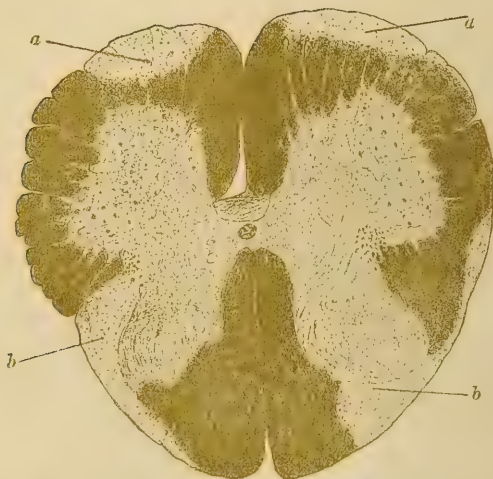


FIG. 156.—Transverse section through the lower part of the lumbar enlargement in a case of cerebro-spinal sclerosis. Stained with osmic acid, mounted in Farrant's solution, and magnified about 8 diameters.

a, a, symmetrical patches of sclerosis in the anterior columns; *b, b*, symmetrical patches of sclerosis in the postero-external and posterior part of the lateral columns.

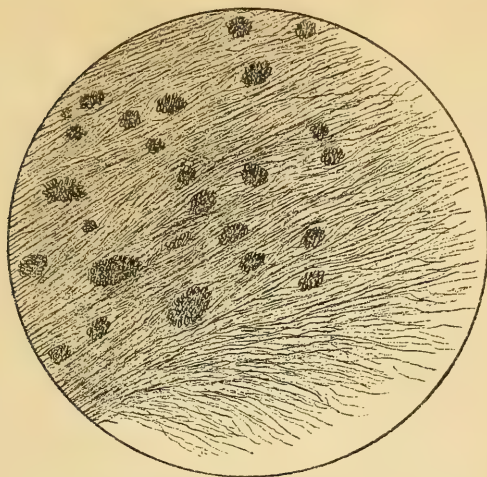


FIG. 157.—*Longitudinal section through a patch of sclerotic tissue in a case of cerebro-spinal sclerosis. Osmic acid and Farrant. $\times 250$ diameters.*

All the nerve tubes have disappeared, and are replaced by bundles of delicate connective tissue. Numerous fatty granules and (?) crystals are scattered throughout the section.

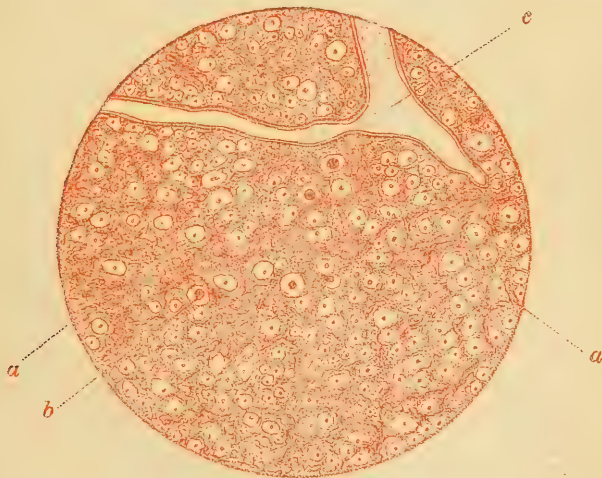


FIG. 158.—*Transverse section through the lateral column of the cord in a case of cerebro-spinal sclerosis, showing the early stage of the lesion. Stained with carmine, mounted in dammar, and magnified about 250 diameters.*

a, a, connective tissue between the transversely divided nerve tubes, which are, in places, widely separated: *b*, hypertrophied axis-cylinder; *c*, longitudinally divided blood-vessel.

The exact cause of the disease is unknown. In some cases, the symptoms seem to have developed after exposure to cold and wet; in others after traumatic injuries (such as blows on the head or spine), mental anxiety and other depressing conditions; in a considerable number of cases, the disease seems to have developed after an attack of pneumonia, typhoid, small-pox, etc. Syphilis does not seem to be a cause of the disease.

The extensive way in which the lesions are diffused throughout the nerve centres is highly suggestive of a toxic cause; while the microscopic characters of the lesion, which seem to show that the condition is a chronic inflammation of the neuroglial tissue, suggest the presence of an irritant. The fact that in a not inconsiderable proportion of cases the disease seems to have followed an acute infectious disease lends support to this view. Marie in particular is a strong advocate of this mode of origin; he states that the sclerotic lesions begin in the vascular walls and develop around the blood-vessels.

The nature of the irritant which is probably the cause of the condition is unknown.

In the present position, then, of our knowledge, it is impossible to say more than this, that the sclerotic lesions are probably the result of an irritant which is distributed through the nerve centres by the blood and blood-vessels; and that the irritant (micro-organism, toxin, ptomaine, or whatever it may happen to be) seems to have a special affinity for the neuroglial tissue of the white matter of the nerve centres and (perhaps) a special destructive action upon the white substance of the nerve tubes. It is difficult, I think, on any other supposition to account for the fact that the grey matter of the cerebral cortex, which is so richly supplied with minute blood-vessels and in which secondary deposits carried by the blood-vessels (secondary deposits of melanotic sarcoma, for example) are often abundantly developed, is so rarely affected (i.e. seat of the sclerotic lesions).

Clinical History.—In the great majority of cases, the onset is slow and insidious. In rare and exceptional cases the symptoms seem to be developed more abruptly; in one of my cases, for example, the affection commenced somewhat suddenly with deafness and noises in the head after exposure to a hot sun.

As we might naturally expect from the multiplicity of the sclerotic lesions and the extremely irregular manner in which they are distributed throughout the nerve centres, the symptoms are multifarious; in fact, as Charcot has pointed out, the affection is a polymorphous one.

In the great majority of cases, the sclerotic patches are situated both in the brain and spinal cord; but in rare cases they are limited to, or chiefly distributed in, the spinal cord or the brain. Three types of the disease have consequently been described, viz.:—(1) The *cerebro-spinal*; (2) The *spinal*; and (3) The *cerebral* types of the disease. The cerebro-spinal form is by far the most common variety; but the spinal and cerebral forms are none the less important, for they comprise the cases in which the diagnosis is attended with the greatest difficulty.

It is unnecessary to say that in the common variety of the disease—the cerebro-spinal form—both cerebral and spinal symptoms are present. In some of these mixed cases, the cerebral symptoms are most conspicuous; while in others, the spinal symptoms are most prominent. This statement applies more particularly to the early stages of the disease.

In some of the mixed (cerebro-spinal) cases, the earlier symptoms of the disease are chiefly or exclusively spinal (muscular weakness, difficulty in walking, a spastic condition of the lower extremities, exaggeration of the knee-jerks, the presence of ankle-clonus, for example). In others, the earlier symptoms of the disease are chiefly or exclusively cerebral (vertigo, impairment of speech, deafness, loss of emotional control, hysterical symptoms, for example).

But notwithstanding these differences of detail and notwithstanding the varied character which the clinical picture may present, well marked and fully developed cases of multiple cerebro-spinal sclerosis are attended with certain outstanding and characteristic symptoms which are quite distinctive. The more important of these symptoms are:—(1) a peculiar form of tremor; (2) a characteristic affection of speech; and (3) nystagmus. Other important and characteristic symptoms are loss of motor power (paresis), exaggeration of the knee-jerks, the presence of ankle-clonus, a silly, vacant expression of countenance and certain ocular derangements (optic atrophy and visual defects). Cerebral symptoms, to which I will presently refer in more detail,

are sometimes developed in the later stages of the disease. In some cases, there is spastic paraplegia, in others inco-ordination.

In the great majority of cases, the sensory functions of the skin are normal or only slightly impaired, the functions of the bladder and rectum are not markedly affected and there are no trophic disturbances. Vasomotor derangements (such as a blue, cold condition of the hands and feet) are occasionally but rarely present; in two or three cases which have come under my notice, they were conspicuous features in the early stages of the case. These (negative) statements chiefly apply to the earlier stages of the disease. In the final stages, sensory disturbances, paralysis of the bladder, cystitis and bed-sores may be developed.

Let us consider some of the more important symptoms in detail.

Loss of motor power ; paralysis.—Motor impairment is, as I have already mentioned, a prominent feature in many cases of the disease; weakness in the legs is often the first symptom which is developed. The motor weakness is incomplete; there is rarely marked paralysis, until at all events the later stages of the case. The motor weakness is apt to vary in degree from time to time; sudden but temporary loss of motor power in one or both legs occasionally occurs.

The condition of the muscles.—The muscles of the affected limbs are usually of normal bulk and their electrical reactions are unaltered. In the advanced stages of the disease, localised muscular atrophy is occasionally though very rarely developed; it usually appears to be due to the extension of a sclerotic patch to the anterior horn of grey matter and destruction of the multipolar nerve cells or their axis-cylinder processes.

The condition of the reflexes.—In the great majority of cases the *deep* reflexes are exaggerated; the knee-jerks are usually markedly increased, even in the earlier stages; as the disease advances, most marked and typical ankle-clonus is often developed. The exaggerated condition of the deep reflexes is an important point of distinction between multiple cerebro-spinal sclerosis and Friedreich's ataxia.

The *skin* reflexes do not present any characteristic alterations; in some cases they are normal; in others diminished; in others exaggerated.

The characteristic tremor.—This is the most important symp-

tom of the disease from a diagnostic point of view. There is no tremor so long as the patient is at perfect rest. The tremor only occurs on voluntary movement. Hence the terms 'volitional' and 'intentional' tremor which have been applied to it. The tremor is rhythmical, coarse and jerking in character. The trembling movements are of considerable amplitude; they gradually increase in size and frequency the longer the muscular movement which initiates them continues; they are said to number six or seven per second. The characters of the tremor are diagrammatically represented in fig. 159. In consequence of the occurrence of this rhythmical jerking tremor, movements

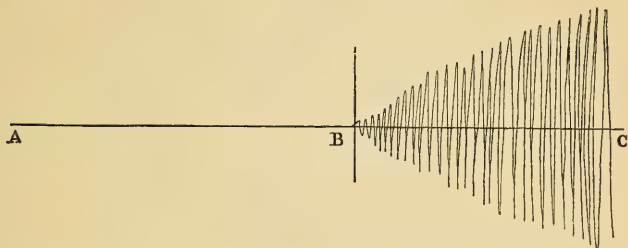


FIG. 159.—*Diagrammatic representation of the tremor in cerebro-spinal sclerosis.*
—(After Charcot.)

During repose (line A to B) there is absolutely no tremor. When the patient begins to make a movement (at B) the tremor commences and continues and increases during the continuance of the movement (B to C).

are rendered extremely unsteady, though their main direction is preserved. The tremor may occur with any voluntary movement, i.e. may affect any of the muscles, but is generally best marked in the upper extremities. Like almost every form of tremor, the characteristic tremor of multiple cerebro-spinal sclerosis is increased and intensified by agitation and excitement.

Even in the advanced stages of the disease and in those cases in which every voluntary effort is attended with marked tremor there is no tremor so long as the patient is lying at absolute rest in bed. It is only when he makes a voluntary movement—speaks, puts out the tongue, or raises the head, arm or leg from the bed—that the tremor is produced.

When the patient is seated in a chair, there may be nothing noticeable, or at most a slight rhythmical jerking of the head. You must remember that in order to maintain the erect position of the head and trunk even in the sitting position, some voluntary effort on the part of the muscles of the back and neck is required; hence there is some tremor and the head and neck shake.

When the patient attempts to carry a glass of water to the mouth, the tremor is exquisitely demonstrated. This is one of the routine tests which we always employ when we suspect the presence of multiple cerebro-spinal sclerosis. The main direction of the movement is preserved and the glass reaches the face; but as it is carried to the mouth it is jerked up and down, the water is violently agitated and, unless the quantity is small, is spilt. When the glass approaches the mouth, the tremor becomes much more marked; the tremor affects the muscles of the head and neck as well as those of the arm, and the head is violently jerked. Finally, the glass may be suddenly and forcibly driven against the teeth, nose or chin and its contents dashed over the patient's face or neck.

Some patients attempt to dodge the tremor, i.e. to reduce the amplitude of the jerking movements of the arm and head, by closely fixing the arm and forearm to the side of the trunk, by throwing the head back or fixing it firmly down between the shoulders, or by firmly grasping with the other hand the wrist of the hand in which the glass is held; but even these manœuvres do not prevent the trembling, they only lessen its range, and the water may still be spilt.

In those cases in which the tremor and unsteadiness of movement are very great, the patient may refuse to make the movement, or if he attempts to do so may dash the glass from the table in his efforts to grasp it.

Even in the early stages of the disease and in those cases in which the tremor is only slightly marked, the tremor can usually be well demonstrated by making the patient write. When the tremor is more marked the patient may be unable to write.

When the patient walks, the head may be seen to jerk in a rhythmical way. In some cases, the lower extremities as well as the body shake.

In the advanced stages of the disease, articulation may be attended with twitchings and tremors in the lips and facial muscles. A rhythmical tremor is sometimes also seen in the tongue when it is protruded.

The characteristic tremor is only developed when the lesion is fairly well advanced (i.e. when the axis-cylinders have been laid bare). It is not present in the earliest stages of the disease; but in the great majority of cases it can be demonstrated when the patient first consults the physician. It is sometimes present in one arm, but not in the other; more often it is merely more marked in one arm than in the other. In two cases which have come under my notice recently, the patient could carry a glass of water to the mouth quite steadily with one hand (i.e. without any tremor); while the characteristic tremor was developed in a typical and marked degree when he performed the same movement with the other.

The cause of the tremor.—Charcot advanced the view, and his opinion is accepted by most authorities, that the tremor is due to irregular conduction of nerve force through axis-cylinders which are, so to speak, lying naked (uninsulated) in the midst of the sclerosed tissue. We have seen that an important pathological feature of the disease is the disappearance of the medullary sheath and the laying bare, as it were, of the axis-cylinder. The white substance of Schwann seems to act as an insulator to the axis-cylinder; when it is removed or destroyed, the conduction through the axis-cylinders becomes irregular, intermittent, jerky and unsteady—just as the conduction of an electric current through an imperfectly insulated copper wire, instead of being smooth and steady, is apt to become irregular, jerky and intermittent. This explanation of the tremor is supported by the fact that a similar form of tremor seems to be produced by pressure on motor nerve tubes. In some cases of cerebellar tumour, for example, voluntary movements in the upper extremities are attended with a rhythmical, jerking tremor, which closely resembles that characteristic of multiple sclerosis and which is probably due to the pressure of the tumour upon the medulla oblongata and interrupted or irregular conduction of voluntary motor impulses (nerve force) through the fibres of the pyramidal tracts.

Charcot's explanation of the mode of production of the

tremor is not accepted by all authorities. Erb, for example, has suggested that it is due to the presence of sclerotic nodules in some special parts of the brain—the pons and adjacent parts of the mesencephalon. The fact that in the early stages of some cases which ultimately present all the characteristic features of multiple cerebro-spinal sclerosis (and in which the lesion is presumably confined to the spinal cord and to the crossed pyramidal tracts) there is no tremor but merely a condition of spastic paraplegia, perhaps lends some support to Erb's opinion. But this argument is not conclusive, for the exact nature of the lesion in the early stages of cases of this description is uncertain. It is possible that in cases of this kind the lesions characteristic of multiple cerebro-spinal sclerosis are superadded to the lesion characteristic of primary sclerosis of the crossed pyramidal tracts.

The gait.—Patients affected with multiple cerebro-spinal sclerosis walk in a variety of ways. The different forms of gait of course depend upon the distribution of the sclerotic lesions, the stage of the disease and the severity of the case.

As I shall afterwards point out when I come to speak of the diagnosis, the clinical picture in the early stages of some rare and exceptional cases is identical with that of primary spastic paraplegia. In cases of this kind, the gait may be typically spastic in character.

When the rhythmical tremor affects the movements of the legs and trunk, the gait is shaky or trembling.

In those cases in which the muscles of the head and neck are alone affected by the tremor, the only alteration which can be detected may be a certain stiffness in the way in which the head is carried and a slight rhythmical jerking of the head when the patient walks.

In those cases in which the movements of the lower extremities are affected in a marked degree, the gait may be extremely unsteady.

In some cases, the gait is partly spastic and partly trembling. As in ordinary spastic paraplegia, the legs are stiff and the feet may appear to be stuck to the ground and to be raised with difficulty, but at each step a rhythmical tremor occurs in the lower extremities and in the trunk, which causes the stiff legs and the body to shake.

In others, the gait is markedly unsteady and inco-ordinate. In these cases, the sclerotic lesions probably implicate the posterior more than the lateral or anterior columns of the cord. In some cases of this kind, the inco-ordination seems chiefly to affect the muscles of the trunk. In some cases, the patient cannot walk deliberately forward in a straight line; he may reel or sway from side to side or may be forcibly propelled forwards.

Speech affection.—This is a highly characteristic symptom of the disease. The articulation is slow and drawling; the individual syllables are pronounced separately (*scanning* speech). The pronunciation of the consonants, such as l, p and g, is usually more defective than that of the vowels. The voice is in many cases high-pitched and singularly monotonous; it sometimes has a nasal twang. In the terminal stages of some cases the speech is unintelligible. In the advanced stages of the disease the voluntary movements of articulation may be attended with rhythmical tremors in the lips, tongue and facial muscles. Rhythmical tremors may also affect the tongue when it is protruded. Like the other motor disturbances, the speech affection is apt to vary in intensity from time to time; indeed, the articulatory defect may, like the paralysis and tremor, disappear for a time; but, speaking generally, the articulation gradually increases as the disease advances.

Facial expression.—In well-marked cases, the facial expression is often highly characteristic; it is vacant, stolid and stupid-looking. The patient has a silly expression of countenance. The appearance of the face is very similar to that which is seen in many cases of Friedreich's ataxia.

LECTURE XXX

MULTIPLE CEREBRO-SPINAL SCLEROSIS (*Continued*)

IN the last lecture, Gentlemen, we were considering the clinical history of multiple cerebro-spinal sclerosis. Let me now refer to some of the ocular defects and alterations which may be present. They are numerous and important.

Nystagmus.—This is a highly characteristic symptom which is present in most cases; it may, however, be entirely absent even in the advanced stages of the disease; there was no nystagmus in a typical and advanced case which recently came under my observation. The nystagmus is almost always horizontal, very rarely vertical. It closely resembles the nystagmus which occurs in Friedreich's ataxia. In the great majority of cases it is only produced when the patient makes a voluntary effort, i.e. turns the eyes to one side or "fixes" an object placed well out to one side. In some cases it is present when the eyeballs are (apparently) at rest. In many cases the nystagmus is probably analogous to the tremor which attends the other voluntary movements. In some cases it is associated with paresis of the ocular muscles.

The condition of the pupils.—The pupils are in some cases normal, in others unequal in size; in other cases markedly contracted (myosis). The activity of the pupil both to light and accommodation may be interfered with, but the Argyll Robertson condition is very rarely if ever present in a typical and fully developed form. Even if the Argyll Robertson condition does occasionally occur, it is probably merely accidental and due to the (accidental) presence of a sclerotic patch in some part (sensory side) of the reflex arc.

Diplopia and paralysis of the ocular muscles.—Temporary diplopia is not uncommon; in a small proportion of cases, temporary paralysis of some one or other of the ocular muscles analogous to that which occurs in locomotor ataxia is developed.

The external rectus is the muscle which is most frequently affected. In some cases the associated movements of the eye-balls, such as convergence, cannot be performed; while the individual movements of each eyeball are uninterfered with.

Optic atrophy.—In a considerable proportion of cases of multiple cerebro-spinal sclerosis, the optic discs present some atrophic changes. In many cases, the degree of atrophy is slight and difficult to detect; this is no doubt the explanation of the very different statements which different observers make as to the frequency with which optic atrophy occurs. If I were to judge from my own experience I should certainly say that optic atrophy, sufficiently well marked to be clearly and definitely recognised by the ophthalmoscope, is only present in a comparatively small proportion of cases. But Buzzard found pallor of the discs in 43 per cent. of his cases; and Uhthoff states that with the exception of cerebral tumour and tubercular meningitis, there is no disease of the nervous system (even including tabes) which is so often accompanied by ophthalmoscopic changes as disseminated sclerosis.

The explanation of this difference of opinion is doubtless that, in common with many other observers who state that optic atrophy only occurs in a comparatively small proportion of cases, I have failed to observe the slighter atrophic changes.

In other cases, the atrophy is well marked. In some cases, the whole disc is atrophied and white; in others, the decoloration is incomplete, the nasal side being comparatively little affected; in a third group of cases, the atrophic changes are confined to the temporal side of the disc, the nasal side being perfectly normal. The atrophic changes may be bilateral or unilateral; they are often much more marked in one eye than in the other.

Diminished acuity of vision; amblyopia.—When the degree of optic atrophy is considerable, the acuity of vision is considerably impaired and the fields of vision (both for white and for colours) are constricted; but these visual defects are often much less marked than one would expect from the degree of optic atrophy which is present. The discrepancy between the functional modifications (the visual defects) and structural changes (the optic atrophy) is probably explained, as Charcot suggested, by the fact that naked axis-cylinders still remain in the optic nerves or optic tracts which are the seat of sclerotic patches.

Even in those cases in which the optic atrophy, as seen by the ophthalmoscope, is very marked, the blindness is very rarely, if ever, complete. *Vice versâ*, there is sometimes marked impairment of vision in cases in which the fundus oculi appears to be quite normal. In such cases it is probable that the sclerotic patches, which are the cause of the dimness of vision, do not extend sufficiently forwards to implicate the disc.

The dimness of vision is in some cases developed slowly and gradually, in others it is rapidly or suddenly established. In some cases, it persists and increases; in other cases it undergoes a gradual, or it may be rapid, improvement; in the latter cases it is apt to return and after a time to become constant.

Constriction of the fields of vision; dyschromatopsia.—In some cases, the fields both for white and for colours are constricted; in others the peripheral vision is normal while the central vision is markedly impaired. In some cases, careful perimeter observations show the presence of scotomata in some part of the field.

Temporary loss of vision.—Temporary loss of vision is in some cases quickly developed and as quickly disappears; it is usually unilateral, but not unfrequently affects first one eye and then the other. It is a most suggestive and characteristic symptom, though, as we shall afterwards see, the fact of its occurrence is apt to lead an inexperienced observer to suppose that he is dealing with a case of hysteria. In some cases, this loss of vision persists instead of being recovered from.

These visual defects are often much more marked in one eye than in the other, or they may be entirely confined to one eye.

Their diverse character and irregular distribution can only be satisfactorily explained by supposing that patches or islets of sclerosis, with varying alterations in the nerve elements, are situated in the optic nerves, optic tracts, optic radiation of Gratiolet, etc.

When I come to speak of the diagnosis, I will refer in more detail to the constriction of the fields and the dyschromatopsia.

Auditory derangements.—In some cases, noises in the ears are complained of; in others the acuity of hearing is diminished; but marked deafness is very rare.

Cerebral symptoms.—Vertigo is, as I have already mentioned, a prominent symptom in many cases of the disease, and it is often one of the earliest symptoms. The giddiness is usually of

momentary or brief duration, very rarely continuous. Headache is occasionally complained of, but in many cases it is altogether absent; in none of the cases which have come under my own notice has it been a prominent symptom. The memory and mental faculties are often decidedly blunted. In many cases the patient becomes irritable and loses his self-control; hysterical symptoms and emotional disturbances are comparatively common; the patient may laugh or cry without any sufficient or obvious cause. In a few rare and exceptional cases, grandiose ideas exactly similar to those which characterise general paralysis of the insane have been present. Illusions and hallucinations and definite symptoms of insanity are rare. Sudden congestive and pseudo-apoplectic attacks, characterised by insensibility, rapid elevation of temperature, quick pulse, and epileptiform convulsions which are sometimes unilateral, occur in a small proportion of cases. These congestive and pseudo-apoplectic attacks are exactly similar to the congestive attacks which are so common in general paralysis of the insane. The patient may die in the attack, but as a rule the coma disappears in the course of a day or two, leaving perhaps behind it a temporary aphasia or a temporary hemiplegia, which in its turn is quickly recovered from.

The condition of the sensory functions of the skin.—The sensory functions are rarely affected in any marked degree. Subjective derangements of sensation, such as numbness, pins and needles, etc., are not unfrequently complained of; but distinct objective disturbances of sensation, such as well-marked anæsthesia and hyperæsthesia, can rarely be demonstrated.

The condition of the bladder and rectum.—The bowels are in some cases constipated. Until the later stages of the disease, the functions of the bladder are rarely interfered with in a marked degree. When the patient becomes bedridden, the bladder may become paralysed and cystitis may be developed; but these complications are infrequent. In the later stages of the disease, the lower extremities are often in a condition of spastic paraplegia; in cases of this kind, marked paralysis of the bladder (with retention requiring the use of the catheter, cystitis, or with incontinence from paralysis of the sphincter) is, as I have already told you, comparatively rare.

Vasomotor and trophic derangements.—As I have already

pointed out, the hands and feet are in some cases blue and cold ; but, speaking generally, vasomotor and trophic derangements are rare.

General state of nutrition and condition of the viscera.—Until the later stages of the disease, the general state of nutrition is usually well preserved and complications on the part of the other systems and organs are rarely developed.

Duration and Course.—In the great majority of cases the course of the disease is essentially chronic. The average duration is probably from eight to twelve years. In rare cases, the disease is much more rapidly fatal. In some cases the duration is longer; the patient may live for fifteen or twenty years. In those cases in which the lesion is, at its commencement, limited to the spinal cord (the spinal type of the disease) the duration is usually longer than in those cases in which the cerebral nerve centres are affected.

As the disease advances, the motor paralysis may become more marked; a complete condition of spastic paraplegia may be developed and the patient may be confined to bed or to his chair. The paralysis is sometimes complete; the legs may be rigidly extended, the deep reflexes increased and the condition exactly resemble the advanced stage of spastic paraplegia. The tremor, too, with the advance of the disease becomes intensified, and attempts at voluntary movement not only produce rigidity and spasm but may cause violent clonic movement of the muscles and forcible jerking and shaking of the whole body. The mental deterioration gradually becomes greater, the speech more scanning, monotonous and weak until ultimately it may become unintelligible.

In the later stages of the disease, the general state of nutrition may be affected. The patient may become emaciated; localised muscular atrophies may, as I have already stated, be produced. Bulbar symptoms—difficulty in swallowing—paralysis of the bladder, cystitis and bedsores may occur. Ultimately the patient dies either from simple exhaustion, cystitis, bedsores, paralysis of the respiratory muscles, or some intercurrent complication, such as pneumonia or phthisis.

Diagnosis.—In fully developed and typical cases, the diagnosis presents no difficulty. The gradual onset and slow

course of the symptoms, with perhaps periods of marked improvement or it may be complete remission of the symptoms, the presence both of spinal and cerebral symptoms and the peculiar character of the symptoms go to form a striking clinical picture which it is impossible to mistake. The symptoms which are especially important and characteristic from a diagnostic point of view are:—The voluntary rhythmical tremor; the speech affection; the nystagmus; the vacant, silly expression of countenance; the vertigo; the exaggeration of the deep reflexes; the loss of motor power, which in the lower extremities is often spastic in type; the ocular alterations; and the absence of any marked impairment of the sensibility of the skin, of the functions of the bladder and rectum, and the absence of vasomotor and trophic disturbances.

In the earlier stages of the disease, in the rare cases in which the symptoms are exclusively spinal or exclusively cerebral, and in the '*fruste*' cases in which the tremor and other characteristic symptoms are absent or are only slightly marked, the diagnosis may be extremely difficult or for a time impossible.

The differential diagnosis of multiple cerebro-spinal sclerosis and of primary spastic paraplegia.—There is no difficulty in distinguishing these affections when the characteristic symptoms of cerebro-spinal sclerosis are fully developed. But cases are occasionally met with in which, in the earlier stages at all events, the diagnosis is impossible. Some fourteen years ago I saw on several occasions with Dr. Sinclair of Dundee a young lady who presented all the characteristic features of primary spastic paraplegia—motor impairment, rigidity and stiffness in the lower extremities, with marked exaggeration of the knee-jerks and ankle-clonus. After some years, the characteristic symptoms of multiple cerebro-spinal sclerosis were gradually developed; and now, the case is a typical and characteristic example of that disease. I may say in passing that in this case the spastic paraplegia followed upon (? and was perhaps due to) arrested menstruation, and that in its early stages symptoms indicative of vasomotor disturbance (coldness, blueness, and swelling of the hands and feet) were prominent.

It is possible that in cases such as this the disease at its commencement is nothing more than a primary sclerosis of the crossed pyramidal tracts, and that at a later stage multiple cerebro-

spinal sclerosis is superadded. But it seems more reasonable to suppose that the case was from the first one of multiple cerebro-spinal sclerosis and that in the early stages the lesions (the sclerotic patches) were confined to the spinal cord, i.e. to the lateral columns.

Several other cases of the same kind have come under my notice, and in dealing with cases which appear to be primary lateral sclerosis in young women, I now always keep in view the possibility of their ultimately turning out to be cases of disseminated sclerosis.

The differential diagnosis of multiple cerebro-spinal sclerosis and locomotor ataxia.—This rarely presents any difficulty, but in some exceptional cases of cerebro-spinal sclerosis the patches of sclerosis appear to be confined to, or to chiefly involve, the posterior columns of the cord. In exceptional cases of this kind, the symptoms of locomotor ataxia are more or less closely simulated, and the diagnosis may for a time be exceedingly difficult or impossible.

The chief points of distinction between locomotor ataxia and disseminated sclerosis are as follows:—

1. *The age.*—Locomotor ataxia is seldom developed before the age of thirty; cerebro-spinal sclerosis is most frequently developed between the ages of twenty and thirty.

2. *The sex.*—Locomotor ataxia is much more frequent in males than in females; while cerebro-spinal sclerosis probably occurs with equal frequency in the two sexes, or is perhaps more common in women.

3. *The condition of the reflexes.*—In locomotor ataxia the knee-jerks are abolished; whereas in the great majority of cases of cerebro-spinal sclerosis the knee-jerks are increased. In locomotor ataxia, the Argyll Robertson condition of the pupil is often present, but in multiple cerebro-spinal sclerosis the Argyll Robertson condition of the pupil is of very rare occurrence. In locomotor ataxia, the rectal, vesical and sexual reflexes are usually deranged; while in multiple cerebro-spinal sclerosis the rectal, vesical and sexual reflexes are usually normal, at all events until the later stages of the case.

4. *The condition of the sensory nerve apparatus.*—Lightning pains, anæsthetic patches and loss of the muscular sense, which are such characteristic features in most cases of locomotor ataxia,

are very rarely met with in cerebro-spinal sclerosis. In those cases in which a sclerotic patch invades the posterior columns of the cord these symptoms may no doubt be present.

5. *The presence of motor paralysis.*—In locomotor ataxia the muscular power is usually well preserved until the terminal stages of the case; but in cerebro-spinal sclerosis some impairment of motor power is generally an early feature.

6. *The presence of other symptoms characteristic of disseminated sclerosis.*—The volitional tremor, the nystagmus, the speech affection, the vertigo, the facial expression and the impairment of the mental faculties, which are such characteristic features of cerebro-spinal sclerosis, do not occur in locomotor ataxia.

Even in those cases of multiple cerebro-spinal sclerosis in which the gait is unsteady from ataxia as well as from muscular weakness and tremor, a careful observer could not mistake the case for one of locomotor ataxia. The inco-ordination is altogether different; it is coarser and it affects the trunk muscles more than those of the legs. The patient sways from side to side or is propelled forwards or from side to side in an inco-ordinate manner. Further, in any doubtful case of this kind the other facts which have been mentioned above (under heads 1 to 6) are quite distinctive and conclusive.

The differential diagnosis of multiple cerebro-spinal sclerosis and cerebellar tumour.—In both conditions there may be headache, nystagmus, giddiness, a reeling gait due to weakness or inco-ordination of the muscles of the trunk, loss of power in the upper and lower extremities, exaggeration of the knee-jerks and the presence of ankle-clonus. Further, it must be remembered that a voluntary tremor, which more or less closely resembles the characteristic tremor of multiple cerebro-spinal sclerosis, is sometimes produced by the pressure of a cerebellar tumour upon the subjacent pyramidal tracts. But notwithstanding these points of resemblance, the two diseases can usually be distinguished without any difficulty. The more important points of distinction are:—

1. *The nature of the symptoms.*—In cerebellar tumour, headache, vomiting and optic neuritis or post-neuritic atrophy are prominent symptoms; whereas in multiple cerebro-spinal sclerosis, headache is usually altogether absent or only slightly marked, vomiting is not present, optic neuritis is most exceptional, and optic atrophy, when it does occur, is primary not post-

neuritic, less intense and very rarely attended with marked or complete blindness.

In cases of cerebellar tumour, the characteristic scanning speech, monotony of voice, and stolid expression of countenance, which are such important features of multiple cerebro-spinal sclerosis, are not present. In the exceptional cases of cerebellar tumour in which a rhythmical tremor on voluntary movement occurs, the tremor is slight in comparison with the motor paralysis; but in multiple cerebro-spinal sclerosis the reverse is the case. In that disease, the volitional tremor is usually much more marked than the motor paralysis.

2. *The age of the patient.*—Cerebellar tumours are most common in children; while multiple cerebro-spinal sclerosis is very rare in childhood. The age of the patient is therefore a point of some importance.

3. *The duration and course of the case.*—In the great majority of cases of cerebellar tumour, the course of the disease is much more rapid and continuously progressive than in cases of cerebro-spinal sclerosis.

The differential diagnosis of multiple cerebro-spinal sclerosis and paralysis agitans.—This presents no difficulty in the great majority of cases, but cases perhaps occasionally occur in which the two conditions are combined. The chief points of distinction are:—

1. *The age of the patient.*—Paralysis agitans is a disease of later adult life and of old age. It rarely occurs before the age of forty-five; whereas, cerebro-spinal sclerosis is essentially a disease of youth and early adult life; it is very rarely developed after the age of forty.

2. *The character of the tremor.*—The movements composing the tremor are less frequent in paralysis agitans than in multiple cerebro-spinal sclerosis. The tremor of paralysis agitans has no resemblance to the tremor of chorea; while the tremor of cerebro-spinal sclerosis does to some extent resemble that of chorea. In paralysis agitans, the tremor is a finer tremor, more rhythmical and more regular than that of cerebro-spinal sclerosis. It occurs while the muscles are at rest, and is often diminished or arrested by voluntary movement. It rarely affects the head, facial and tongue muscles; whereas in cerebro-spinal sclerosis the tremor only occurs on voluntary movement and it often affects the head, facial and tongue muscles.

3. *The associated symptoms.*—In paralysis agitans, the slow scanning speech, nystagmus, vertigo, symptoms of mental impairment, optic atrophy and the other ocular defects which have been described as occurring in a considerable proportion of cases of cerebro-spinal sclerosis are not observed.

The differential diagnosis of multiple cerebro-spinal sclerosis and Friedreich's ataxia.—This has been fully considered in connection with Friedreich's ataxia (see page 380).

The differential diagnosis of multiple cerebro-spinal sclerosis and general paralysis of the insane.—There is no difficulty in distinguishing the two conditions in the earlier stages and if the course and progress of the disease are taken into account; but in the advanced stages of cerebro-spinal sclerosis the symptoms may closely simulate those of general paralysis of the insane. The vacant silly expression of countenance, the speech defect which when advanced is very similar in character in the two diseases, the occurrence of voluntary tremors in the lip, facial and tongue muscles during efforts of articulation, the obvious mental impairment and the enfeebled motor power may throw the observer off his guard and lead him to suppose that he is dealing with a case of general paralysis of the insane. A few months ago, I saw, with Dr. Johnston of Leith, a patient who was suffering from cerebro-spinal sclerosis in an advanced stage. At the first glance, the case appeared to be one of general paralysis. Efforts at articulation were associated with marked tremor in the facial, lip and tongue muscles, the speech was identical with that characteristic of general paralysis in an advanced stage, and the patient had the same vacant dull expression of countenance which is seen in general paralysis; but the previous history of the case, and a very cursory examination, at once showed the true nature of the condition. The disease was of long standing, the patient was a young man, the history showed that in the earlier stages the symptoms had been quite characteristic of multiple cerebro-spinal sclerosis; the legs were in a condition of spastic paraplegia, and movements of the head, neck and arms were attended with the most marked and characteristic volitional tremor.

Further, it must be remembered that in rare cases of multiple cerebro-spinal sclerosis grandiose ideas and congestive or pseudo-apoplectic attacks exactly resembling the congestive attacks which

are so characteristic of general paralysis may be developed. In exceptional cases of this kind, the difficulty of diagnosis is of course greatly increased.

The chief points of distinction are:—

1. *The age of the patient.*—General paralysis of the insane is rarely developed before the age of thirty, while disseminated sclerosis is usually developed between the ages of twenty and thirty.

2. *The sex of the patient.*—General paralysis is rare in women; while multiple cerebro-spinal sclerosis is quite as common in women as in men.

3. *The duration and course of the disease.*—In general paralysis of the insane, the course of the disease is usually much more rapid and the fatal termination is much more quickly reached than in multiple cerebro-spinal sclerosis.

4. *The nature of the symptoms.*—In general paralysis of the insane, the mental symptoms are usually much more conspicuous than the motor (physical) symptoms; whereas in cerebro-spinal sclerosis the reverse is the case. It is only in very rare and exceptional cases of multiple cerebro-spinal sclerosis that the grandiose ideas and marked mental defects, which are so characteristic of general paralysis, are developed.

In general paralysis of the insane, the tremor is a much finer tremor than that which characterises multiple cerebro-spinal sclerosis.

It is unnecessary to enter into further details. It is only in the later stages and in the quite exceptional cases of multiple cerebro-spinal sclerosis in which grandiose ideas and congestive attacks occur that any real difficulty in diagnosis is likely to arise. In advanced cases of this kind, in which the patients are usually bedridden, the lower extremities are often in a condition of marked spastic paraplegia (a condition which is very rarely if ever present in uncomplicated cases of general paralysis of the insane). Further, voluntary movements of the arms, head, trunk, etc., are immediately attended with the production of the characteristic tremor, which at once shows the true nature of the case.

The differential diagnosis of multiple cerebro-spinal sclerosis and of hysteria.—This is sometimes extremely difficult; several cases of cerebro-spinal sclerosis have come under my notice in which a

diagnosis of hysteria had been made, even by experienced and able practitioners.

In the fully developed cases of cerebro-spinal sclerosis which pursue a progressive course, there is no difficulty. But in some of the irregular forms the difficulty is very great. We have seen that multiple cerebro-spinal sclerosis frequently occurs in young women; that in a considerable proportion of cases there is loss of emotional control or more decided hysterical symptoms; and that remarkable periods of improvement and remission are by no means uncommon. Further, in some cases the symptoms have developed suddenly after a moral shock, mental agitation and the like; in others, the vision in one eye is suddenly lost and as suddenly regained. Further, in some hysterical cases the deep reflexes are exaggerated, though in my experience typical ankle-clonus is very rarely if ever developed; and in some cases of hysteria there is tremor.

It is, in short, in those cases of multiple cerebro-spinal sclerosis in which the patient is a young woman, in which hysterical symptoms and emotional manifestations form part of the clinical picture, in which the characteristic tremor and speech affection are only slightly marked, and in which periods of marked remission and improvement in the symptoms (paralysis, amblyopia, etc.) that the chief difficulties in diagnosis occur. Dr. Buzzard, who has directed special attention to this subject, and to whose observations we are greatly indebted, classes these cases under a distinct type—the *hysterical type* of the disease.

He says:—‘But there is a form which is of extreme importance, especially in reference to the subject under discussion, from the fact that the patient is regarded perhaps for years as an example of “hysteria,” and the serious organic disease underlying the hysterical symptoms is very frequently indeed overlooked. In this, which usually, but not invariably, affects the female sex, there is often a history of moral or physical shock, followed by loss of power in one or more limbs, aphonia, convulsive seizures, and alteration of manner of the kind which is termed hysterical.¹ In this form of disseminated

¹ Dr. Buzzard adds the following note:—‘The subject is discussed in my work *On the Simulation of Hysteria by Organic Disease of the Nervous System*. It is not easy to over-estimate the number of cases of this kind which are mistaken for examples of hysteria.’

sclerosis, the characteristic symptoms are usually but slightly marked, even though many years may have passed since the patient first complained of illness. Long continued intervals of apparent recovery are common. Loss of power in one or other of the lower extremities almost always figures in the history, and sometimes this is the only symptom. After an interval of exemption a recurrence of this has taken place immediately after a severe moral shock, and it is doubtless to this circumstance, coupled with the tendency in these cases to emotional manifestations, that the frequency of errors in diagnosis is due.¹

In trying to come to a correct conclusion and to arrive at a definite diagnosis in difficult cases of this kind, the main object of the physician is to determine whether there are any positive indications of organic disease (i.e. of multiple cerebro-spinal sclerosis) which show that there is something more than a mere functional (hysterical) condition. The symptoms which are suggestive or indicative of organic disease (multiple cerebro-spinal sclerosis) are:—

Well-marked volitional tremor; the characteristic speech alterations; nystagmus; marked exaggeration of the deep reflexes and especially the presence of typical ankle-clonus; atrophy of the discs; and certain other ocular symptoms.

When all of these indications are present there is of course no difficulty or doubt; but in those cases in which some of them or only one of them are present there is more room for difficulty or doubt.

Nystagmus is rarely if ever met with in mere hysteria, but is present in a considerable proportion of cases of multiple cerebro-spinal sclerosis.

In hysteria, the knee-jerks are often exaggerated and a spurious or imperfectly developed form of ankle-clonus is not uncommon; but *fully developed and typical ankle-clonus* is, in my experience, rarely if ever observed. In any cases, then, in which the knee-jerks were greatly exaggerated and typical ankle-clonus was present, I should be strongly disposed to suspect the presence of organic disease (multiple cerebro-spinal sclerosis) rather than mere hysteria.

Optic atrophy.—Pallor of the discs and some degree of optic

¹ *British Medical Journal*, vol. ii. 1893, page 780.

atrophy is present in a considerable proportion of cases of multiple cerebro-spinal sclerosis, but is probably never present in mere hysteria. Dr. Buzzard states that in the cases of disseminated sclerosis of the hysterical type which he examined, atrophic changes in the disc were present in no less than 46·6 per cent. In all doubtful cases the condition of the optic discs should be very carefully examined.

The condition of the fields of vision; dyschromatopsia; and amblyopia without visible changes in the fundus.—The visual fields may be contracted, and amblyopia and dyschromatopsia may be present both in multiple cerebro-spinal sclerosis and in hysteria. But according to Buzzard and Head the exact character of these visual alterations is different in the two conditions; they state that in multiple cerebro-spinal sclerosis the fields for white and colours (red, green, etc.) are contracted, but between the limits of the field for white and the fields for red and green an intermediate zone exists in which red appears as black and green as white; while in those cases of hysteria in which the fields for white and colour are markedly contracted, this intermediate zone (in which red is seen as black and green as white) is not present.

Further, Dr. Head has found that in those cases of disseminated sclerosis with so-called hysterical manifestations in which there are no visible (atrophic) changes in the discs, a similar intermediate zone exists. Dr. Buzzard therefore states that the presence of a contracted field of this type should make us hesitate to diagnose 'hysteria' and that it strongly points to a functional change in the nerve which may ultimately end in atrophy.

Transitory blindness; temporary amblyopia.—As I have already pointed out, temporary blindness or amblyopia, usually only affecting one eye, is occasionally met with in cases of multiple cerebro-spinal sclerosis; it may be unattended with any visible changes in the fundus; and if associated with other symptoms suggestive of hysteria (as it usually is in cases of multiple cerebro-spinal sclerosis of the hysterical type) it may be thought to confirm the diagnosis of hysteria.

I have seen more than one case which fully confirms Dr. Buzzard's statement on this point. He says: 'The visual troubles are very frequently themselves a fertile source of error. A girl is brought to an ophthalmologist because she has more or

less suddenly lost the sight of one eye. It is probable that the ophthalmoscope fails to detect the slightest sign of any change in the fundus. The girl, on inquiry, is said to be emotional, and on some previous occasion to have partially lost the power of a limb, which, however, had been absolutely recovered. Or there may be a history of numbness or deadness in a limb. These are all symptoms which, it is notorious, have been looked upon as characteristic of hysteria. I do not know whether such is the case; indeed I have grave doubts upon the point; but I am sure, from observation, often extending over many years, of a large number of cases, that they are symptoms of a stage of disseminated sclerosis. I venture to urge this more particularly because it is quite possible that the patient, when seen by the ophthalmologist, may present as little objective symptoms in her body generally as she does in the eyes. The reflexes may be found, perhaps, somewhat exaggerated, as they usually are in hysteria, and nothing else may be noticeable. The unilateral blindness in these circumstances is very likely indeed to be explained as "hysterical"; the patient is informed that she will certainly recover her sight, and she and her friends go away rejoicing. In a few weeks, or possibly months, the sight gradually returns. After an uncertain interval, a similar condition may recur, either in the same or the other eye. Again, no decoloration of the disc is perceptible, and the diagnosis of "hysteria" appears to be confirmed. Let the case be followed up, and it will be found that gradually very unmistakable symptoms of disseminated sclerosis will appear, and one or both of the optic discs will be found to be atrophic.¹

From these statements you will understand that the differential diagnosis of multiple cerebro-spinal sclerosis and hysteria is in some cases attended with great difficulty; but that a correct opinion can generally be formed if the whole circumstances of the case (mode of development, nature of the symptoms, progress and course) are carefully and judicially considered.

Prognosis.—The prognosis of multiple cerebro-spinal sclerosis is extremely unfavourable. With perhaps very rare exceptions—for a few cases have been recorded in which recovery is said to have taken place—the disease sooner or later terminates in

¹ *British Medical Journal*, vol. ii., 1893, page 783.

death. In the great majority of cases the course is very slow and chronic. In some cases, the symptoms, once they are developed, pursue a gradually progressive course; but in others, the downward progress is from time to time interrupted by periods of remarkable improvement or it may be temporary remission and disappearance of the symptoms. These remissions are a frequent source of error in diagnosis and prognosis; they are extremely apt to throw the observer off his guard, to suggest hysteria and to lead the practitioner to suppose that the case is going to terminate favourably.

The prognosis as regards duration is more favourable in those cases in which the lesions are confined to, or chiefly situated in, the spinal cord.

The development of well-marked bulbar symptoms, of congestive or pseudo-apoplectic attacks, of emaciation, paralysis of the bladder, cystitis, bed-sores and respiratory or other complications shows that a fatal termination will in all probability occur before long.

Treatment.—We know of no drug which exerts any specific influence upon the course of the disease, though nitrate of silver and arsenic in some cases appear to be attended with temporary benefit. In those cases in which spastic symptoms are prominent, hydrobromic acid or bromide of potassium are sometimes, I believe, useful. In other cases in which the symptoms are chiefly paralytic, strychnine or nux vomica may be given. Iodide of potassium has also been recommended, but in none of the cases in which I have prescribed it has it appeared to be of the slightest use.

The general health should be kept in the best possible state of efficiency and the patient carefully protected from overfatigue and anything which is likely to hasten the development of the disease or to produce respiratory or other complications. It is unnecessary to say that he should be well clothed, well housed and well fed.

Massage, electricity, hydropathy and suspension have been recommended by some authorities; but it is doubtful if they produce any beneficial effect upon the course of the disease.

LECTURE XXXI

SPINAL MENINGITIS

Introductory remarks; relationship of the spinal membranes to the cord and nerve roots.—To-day, Gentlemen, I propose to commence the consideration of spinal meningitis; but before doing so let me briefly direct your attention to the relationship of the spinal membranes to the cord and nerve roots.

The *spinal dura mater* is a dense tough membrane, which is loosely attached externally to the bones of the spinal canal, being separated from the periosteum by loose areolar tissue, fat cells and blood-vessels, and which forms a loose covering or sheath for the spinal cord and its nerve roots, surrounded by the pia, arachnoid, and spinal fluid. (See fig. 160.) Opposite each intervertebral foramen the dura-mater theca presents two openings, placed side by side, which give passage to the two roots of the corresponding spinal nerves. It is continued as a tubular projection on the nerve (fig. 161), and is lost in its sheath. Besides this it is connected with the circumference of the foramen by areolar tissue.¹

The *spinal pia mater* is thicker, finer, and less vascular than the pia mater of the brain, and consists of two layers, viz., a thick external layer consisting of longitudinal bundles of fibres, and an inner thinner layer which is closely applied to the surface of the spinal cord. The two (outer and inner) layers are separated here and there by lymphatic spaces, and between them, the blood-vessels of the pia, which are numerous, are situated. A thick fold of the pia passes down into the anterior median fissure, and a thinner process forms the posterior median septum. From the pia mater, numerous blood-vessels (each covered with a lymphatic sheath) pass into the substance of the cord, and are distributed both to the grey and to the white matter.

¹ Quain's *Anatomy*, ninth edition, vol. ii. p. 372.

The *spinal arachnoid* is an extremely delicate membrane, which forms a very loose covering for the spinal cord. The arachnoid is separated from the dura on the one hand by a space

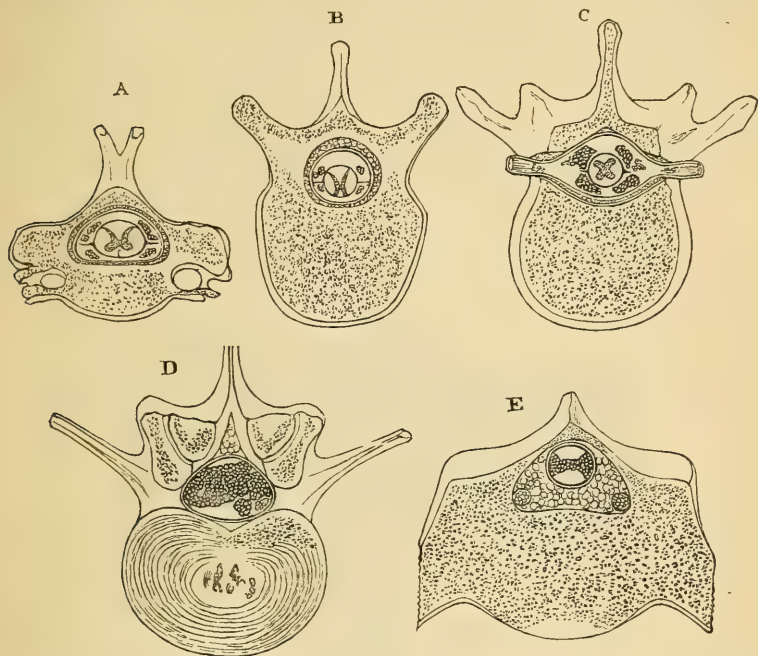


FIG. 160.—Sections showing the general relations of the spinal cord to the inclosing theca, and of this to the vertebral canal.—(Key and Retzius.)

A, section through the fifth cervical vertebra; B, through the tenth dorsal vertebra; C, through the first lumbar vertebra and the foramen of exit of the twelfth dorsal nerve-roots; D, through the disk between the second and third lumbar vertebrae; E, through the first sacral vertebra. In A, B, and C, the cord, covered by pia mater, is seen in the centre, with the ligamentum denticulatum attached to it on either side; the nerve-roots on either side form small groups which, since they pass obliquely downwards to their foramina of exit, are cut across; the dura mater sheath is separated by a considerable space from the cord, and by a quantity of loose areolar and fatty tissue from the wall of the vertebral canal. This tissue is in smaller amount in C. D and E are below the termination of the cord, and show sections of the nerve-bundles of the cauda equina within the theca, which is very large in D, but comparatively small in E, the vertebral canal in the latter being largely occupied by adipose tissue. In this are seen the sections of the two large veins. The arachnoid is not represented in any of these sections.

which is called the *sub-dural space*, and from the pia mater on the other by the *sub-arachnoid space* (see fig. 162).

The sub-arachnoid space is not a continuous space, but is split

up by the *ligamentum denticulatum* and the *septum posticum*. Through this sub-arachnoid space the anterior and posterior nerve roots pass in making their way from the spinal cord on the one hand to the dura mater on the other.

There are two chief forms of spinal meningitis. In the first, the inflammation commences in, or is chiefly or exclusively limited to, the pia mater and arachnoid—the soft membranes; this form we term *leptomeningitis*. In the second, the inflammation commences in, or is chiefly or exclusively confined to,

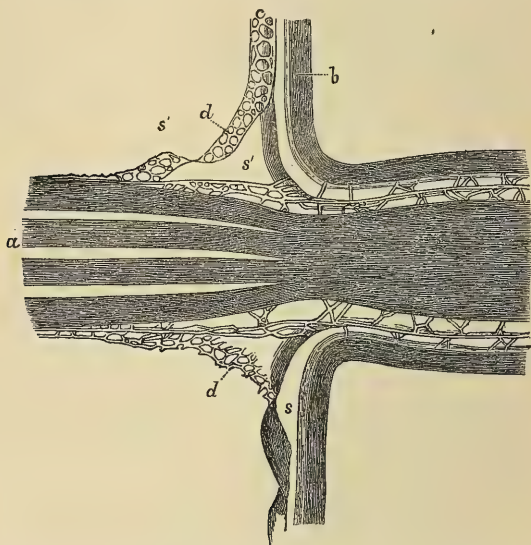


FIG. 161.—Section through the place of exit of a spinal nerve root through the dura mater.
—(Key and Retzius.)

a, bundles of the nerve root becoming collected into a single bundle as they emerge; *b*, dura mater; *c*, arachnoid; *d*, a reticular lamellæ of the arachnoid reflected along nerve root; *s*, sub-dural space; *s'*, *s'*, sub-arachnoid space.

the dura mater or hard membrane; this form we term *pachymeningitis*.

In many cases the two forms (leptomeningitis and pachymeningitis) are combined. In acute inflammation of the pia mater and arachnoid (acute leptomeningitis) the internal surface of the dura is almost always implicated; and in some cases in which the inflammation commences on the outer surface of the dura, the soft membranes ultimately become affected. This is, how-

ever, exceptional. It does, however, occur in some cases of Pott's disease of the vertebræ. In the rare condition which is termed pachymeningitis cervicalis hypertrophica, a form of inflammation to which I shall afterwards have to refer in more detail, the dura mater becomes very much thickened and incorporated, as it were, in one continuous mass with the soft membranes and the cord itself.

Spinal meningitis may be acute, subacute, or chronic; it may be general or localised; it may be primary or secondary. Acute

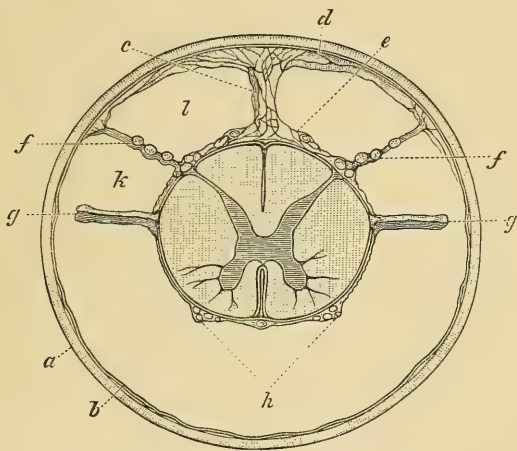


FIG. 162.—Section through the spinal cord and its membranes in the upper dorsal region.
—(After Key and Retzius.) Magnified.

a, dura mater; *b*, arachnoid; *c*, septum posticum; *d*, *e*, *f*, sub-arachnoid trabeculæ, those at *f*, *f*, supporting bundles of a posterior nerve root; *g*, ligamentum denticulatum; *h*, sections of bundles of an anterior nerve root; *k*, *l*, sub-arachnoid space.

inflammation of the membranes of the spinal cord is less common than acute inflammation of the cord itself (acute myelitis), and is much less frequent than acute inflammation of the membranes of the brain (acute cerebral meningitis); nevertheless, it is an important affection. The inflammatory affections of the spinal membranes which are primary, generalised and acute usually affect the soft membranes; whereas, the inflammations which commence in the dura mater are usually localised, subacute or chronic. Acute generalised pachymeningitis does, it is said, occur; but it is so exceedingly rare that for practical purposes it may be ignored. The acute generalised inflammation of the soft membranes (acute

generalised leptomeningitis) is by far the most important form of acute spinal meningitis ; whereas, the chronic localised inflammation of the dura mater is the most common and important form of chronic spinal meningitis. Chronic inflammation of the soft membranes (chronic leptomeningitis) no doubt occurs ; but we know comparatively little about it clinically. It is difficult to diagnose, for the symptoms to which it gives rise are in most cases indefinite and obscure. It is important to remember that localised inflammations of the hard membranes are in most cases secondary, usually the result of some disease of the vertebræ and especially of caries (Pott's disease); consequently, in cases of chronic localised pachymeningitis the clinical picture is usually composed partly of symptoms due to the inflammatory condition of the membranes, and partly of symptoms the result of the primary disease on which the inflammation of the dura mater depends.

In considering the subject of spinal meningitis, I propose to take acute leptomeningitis and chronic pachymeningitis due to Pott's disease as types of the acute and chronic forms of inflammation respectively.

ACUTE LEPTOMENINGITIS

Acute inflammation of the soft membranes is, as I have already stated, in the great majority of cases more or less generalised ; that is to say, it affects the whole or the greater part of the soft membranes within the spinal canal, although the inflammation is, of course, in many cases more marked at some parts than at others.

Morbid Anatomy.—The appearances differ very considerably in different cases, and depend *firstly*, upon the stage of the disease at which the case is examined by the pathologist ; and *secondly*, upon the pathological character or variety of the inflammation, whether sero-fibrinous, purulent, tubercular, etc.

In the early stages (but it is seldom, except in the tubercular form, that the opportunity occurs of examining the beginning of the process, for spinal meningitis is rarely fatal in the early periods of the disease) the chief appearances are those of inflam-

matory congestion. The vascularity of the membranes is increased and the vessels of the pia mater in particular are engorged with blood. The inner surface of the dura has lost its normal smooth polish and presents a somewhat velvety appearance due to commencing swelling and exudation. Small extravasations of blood are in some cases present. The spinal fluid is usually, even at this early stage, somewhat increased in amount and slightly turbid.

In the more advanced stages, the chief appearances are those of inflammatory exudation. The spinal fluid is still more abundant and more turbid; it now contains flakes or flocculi of lymph; in some cases it looks as if it were semi-purulent; in others, the membranes and the surface of the spinal cord are bathed in pus. The inflammatory exudation, whether it consists of lymph or pus, is usually most abundant on the posterior surface of the cord, and at the lower end of the spinal canal, for patients affected with spinal meningitis usually lie on the back, and the inflammatory fluid and exudation products, of course, gravitate to the most dependent part of the spinal canal. The nerve strands of the cauda equina are in some, especially the purulent, cases surrounded by nodular thickenings, due to collections of lymph and pus. In tubercular cases, the amount of lymph is sometimes very scanty; in others considerable. The tubercles (grey granulations) can sometimes be seen with the naked eye. In some cases, they are so numerous, especially over the lower end of the cord and the nerve strands of the cauda equina, that the inflamed membrane looks as if it had been dusted over with little grains or particles of sand. But in many of the tubercular cases, there is little to be seen with the naked eye. I have repeatedly detected tubercles in the spinal membranes with the microscope when I had failed to detect their presence with the naked eye. In several of these cases, there were no symptoms suggestive of spinal meningitis during life. I will refer to this point in more detail in connection with the clinical history of the tubercular form of cerebral meningitis. In that condition, in which the spinal membranes are often involved, the inflammation of the cerebral membranes is apt to overshadow the spinal meningitis. In most of these combined cases, the inflammation of the spinal membranes is a trivial condition in comparison with the inflammation of the cerebral

membranes, to which the symptoms and fatal termination are for the most part due.

Further, it is well to remember that in many of the cases in which the inflammation of the spinal membranes is of the purulent form, the membranes of the brain are also affected. In some of these cases of purulent cerebro-spinal meningitis, as for example in the epidemic form of the disease, the inflammation is the result of a septic irritant which is probably distributed through the blood-vessels. In other cases, a purulent inflammation of the cerebral and spinal membranes is the result of a local condition, such as an abscess of the ear which bursts into the cranial cavity and produces a very severe and diffused form of purulent cerebral meningitis, to which secondary inflammation (purulent) of the spinal membranes may be superadded.

In the third or final stage of the disease—that of the absorption of the inflammatory exudation and of the inflammatory products—the chief alterations are adhesions and thickenings of the spinal membranes to one another and to the surface of the cord. But these appearances are comparatively rarely seen in cases of *acute* generalised spinal meningitis; the condition usually proves fatal before adhesions are developed. In the localised, subacute and chronic varieties of the disease, adhesions and cicatricial thickenings are, of course, more frequent.

I need not go into further detail with regard to the morbid anatomy. The points to which I have directed your attention are those which are of chief importance to the practical physician.

Let us now turn to the pathological physiology.

Pathological Physiology.—In order to understand the clinical symptoms which characterise spinal meningitis, it is essential to remember:—

Firstly, that the membranes of the spinal cord, like the membranes and serous coverings of the other organs of the body, are richly supplied with sensory nerves; and that, when the spinal membranes are inflamed, these sensory nerves are irritated, with the result that pain referred to the seat of inflammation is experienced by the patient.

Secondly, that the soft membranes surround and closely cover the nerve roots; consequently, when the spinal membranes

are inflamed, the nerve roots are apt to be (secondarily) invaded, irritated and pressed upon.

Now, *irritation of a sensory nerve root* as it passes through the inflamed membranes may produce:—(a) pain and hyperæsthesia in the area of distribution of the affected nerve, i.e. in the peripheral parts of the body to which that nerve root is distributed; and (b) reflex muscular spasm, more especially in the muscle or muscles to which the anterior nerve roots of the spinal segment, with which the irritated posterior nerve root is connected, are distributed; while *destruction of a posterior nerve root*, or rather interruption of its function by the pressure of inflammatory products, will be apt to produce (a) anæsthesia in the area of distribution of the affected nerve; and (b) diminished or abolished reflex action in the muscle or muscles to which the anterior nerve roots of the spinal segment, with which the affected posterior nerve root is connected, are distributed.

Again, *irritation of an anterior nerve root* will be apt to produce spasms and cramps in the muscles to which it is distributed; while destruction of, or interruption of the function of, an anterior nerve root, will cause paralysis and arrested reflex action.

These considerations are of great importance, for they explain the manner in which many of the symptoms of spinal meningitis are produced.

These *root-symptoms*, as they are termed, occur both in the acute generalised and the subacute and chronic localised forms of meningitis. In acute leptomeningitis, the root-symptoms are apt to be widely distributed, for many nerve roots are usually implicated; whereas, in localised pachymeningitis, the root symptoms are often limited to the areas of distribution of one or more nerve roots. Again, in acute leptomeningitis the root-symptoms, though more widely distributed over the surface and muscular area of the body, are usually less marked than in localised pachymeningitis: for the nerve roots as they pass through the inflamed hard membrane and the massive inflammatory products which in that condition are usually present at the seat of the inflammation are, as a rule, more seriously and severely damaged than in cases of acute leptomeningitis in which the affection of the nerve roots is less intense, or, to speak somewhat figuratively, more microscopic.

The same statements apply, as we shall afterwards see, to cerebral meningitis; some of the most important symptoms of

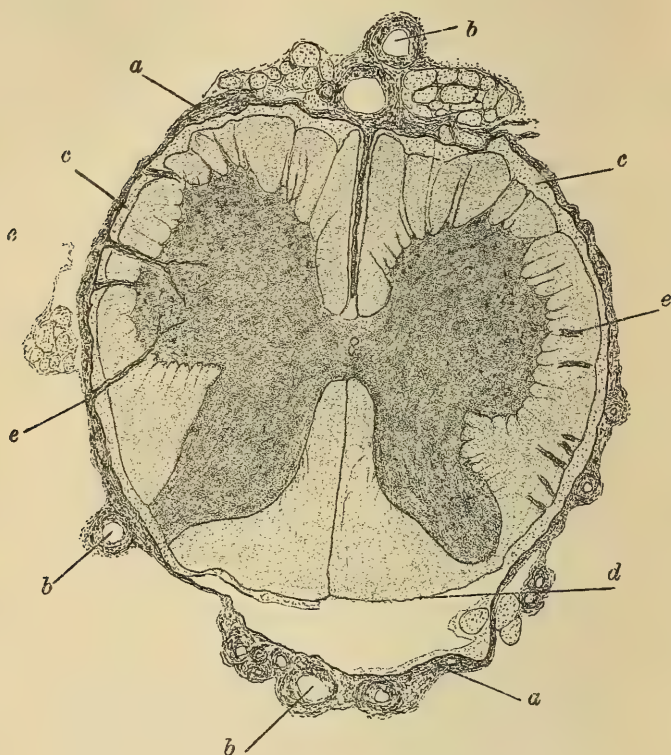


FIG. 163.—*Transverse section through the lumbar enlargement of the cord, showing tubercular disease of the pia mater, and a zone of peripheral myelitis. (\times about 8 diameters.)*

a, a, thickened membranes; *b, b, b*, blood-vessels surrounded with small celled infiltration; *c, c*, the superficial layer of the cord which is inflamed; at *d*, the superficial layer of myelitis has become detached with the pia mater; *e, e, e*, dilated blood-vessels passing from the inflamed membranes into the cord; their coats are, in many instances, surrounded with leucocytes, which cannot of course be seen with the lower magnifying power.

cerebral meningitis are due to irritation of, or pressure upon, the nerves at the base of the brain.

Thirdly, that in cases of leptomeningitis the superficial parts

of the cord are usually implicated. This also is a very important point. It is of course just what would be expected when it is remembered that the pia mater is the vascular membrane of the cord and that innumerable small vessels pass from it into the substance of the cord. When the pia mater is inflamed, the exudation products make their way into the cord substance along the lymphatic sheaths which surround the blood-vessels. In

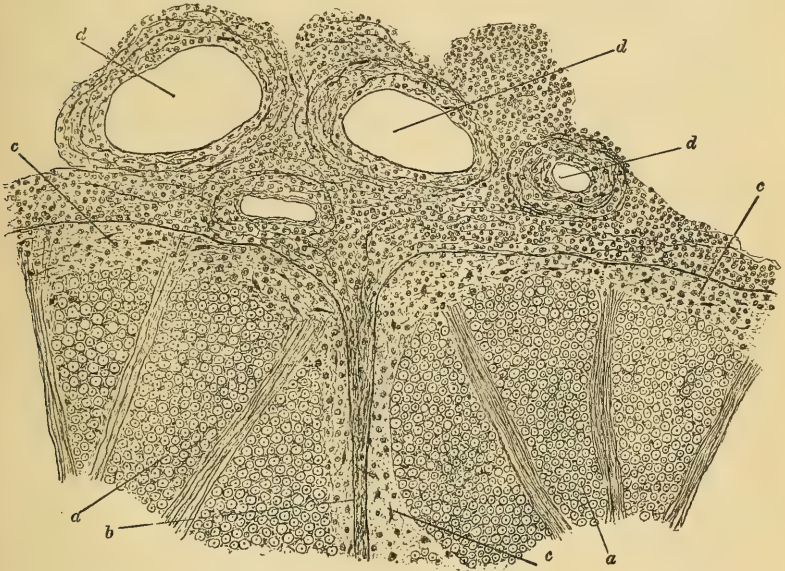


FIG. 164.—*Transverse section through the lower part of the lumbar region of the spinal cord, showing tubercular disease of the pia mater, and a zone of peripheral myelitis. (× about 150 diameters.)*

a, a, anterior columns with bundles of the anterior nerve roots passing through them ; *b*, anterior median fissure filled up with the swollen and inflamed process of pia mater ; *c, c, c*, peripheral layer of myelitis, consisting of the layer of neuroglia, which is situated immediately beneath the pia ; it is much thickened and infiltrated with leucocytes ; *d, d, d*, blood-vessels surrounded with dense masses of small cells, *i.e.* tubercles in an early stage of development.

many cases of leptomeningitis, a peripheral ring of myelitis is present on the surface of the spinal cord. On microscopical examination, the minute vessels which pass into the cord from the pia mater are seen to be dilated and their lymphatic sheaths studded with leucocytes. (See figs. 163 and 164.)

In some cases, the myelitis is more considerable and invades

the deeper parts of the cord. To these cases the term meningo-myelitis is especially applicable.

Exactly the same pathological process occurs in cases of cerebral meningitis; indeed in that condition the symptoms are largely due to implication of the superficial layers of the cerebral cortex, pons Varolii, medulla oblongata, etc.

I would beg you to note that most of the nervous symptoms which occur in spinal meningitis are the result of one or other of the two conditions to which I have just directed your attention, viz., (a) implication of the nerve roots as they pass into or out of the cord, and (b) implication of the cord tissue itself. Leaving out of account the root symptoms and the cord symptoms, pain in the back is the only symptom which is the direct result of the inflammation of the spinal membranes *per se*.¹

Etiology.—The same conditions which produce acute myelitis may produce acute spinal meningitis; but acute primary spinal meningitis is a much rarer condition than acute primary myelitis.

Acute generalised meningitis is probably more common in children and young adults than in older people. This is certainly true of the tubercular variety which is the commonest form of the disease. In adult cases, males are more frequently affected than females.

Acute simple (non-tubercular) leptomeningitis—a very rare condition—seems in some cases to be due to exposure to cold and wet; in others to traumatic injury—I refer, of course, to those cases in which there is no fracture or dislocation of the vertebræ. Spinal meningitis and myelitis the result of traumatic injuries which cause fracture or dislocation are, of course, common conditions; but acute inflammation of the spinal cord and its membranes seems very rarely to result from blows on the back unless the bony projecting covering of the cord is directly and seriously damaged.

The sero-fibrinous form of spinal meningitis, then (if we exclude the tubercular cases), seems usually to be the result of exposure to cold and wet—sleeping out, for example, on damp grass, especially when the resisting power is lowered by an overdose of alcohol.

¹ Irritation of the sensory nerves in the inflamed membranes probably aids (reflexly) in the production of the muscular rigidity and spasm.

The purulent form usually occurs in the course of some septic disease. The epidemic variety (epidemic cerebro-spinal meningitis), in which the exudation is usually purulent, is due to the introduction into the body of a germ poison. Spinal, like cerebral meningitis, is occasionally developed in connection with pneumonia. As I have already remarked, a purulent inflammation of the spinal membranes is sometimes secondary to a purulent inflammation of the membranes of the brain. In the tubercular form this association is common. In rare cases, acute generalised leptomeningitis is secondary to a localised pachymeningitis. The inflammatory process produced on the outer surface of the dura mater by Pott's disease of the vertebræ, for example, may extend through the dura mater and may ultimately involve the soft membranes. This is not common, for the firm dense dura mater is an extremely resisting structure. In the majority of cases of Pott's disease of the vertebræ in which paraplegia is developed, the paralysis is due to compression of the spinal cord by the exudation products on the outer surface of the dura or to myelitis the result of that compression. In many of these cases there is no leptomeningitis. In rare cases, spinal meningitis results from the bursting of an abscess, deep ulcer or bed-sore into the spinal canal.

Clinical History.—The mode of development, rapidity and course of different cases of spinal meningitis is very variable. In some cases of acute generalised meningitis especially in the purulent form, the disease begins abruptly, often with a rigor, is attended with high fever, and runs through a rapid course. In others, the tubercular cases for example, the onset is more gradual and the course more protracted. In some cases, the attack is ushered in by well-marked premonitory symptoms such as pain in the back, headache, vomiting, malaise, slight febrile disturbance, etc. In those cases in which the spinal meningitis is secondary to, or accompanied by, cerebral meningitis, the cerebral symptoms usually occupy the chief place in the clinical picture. In cases of this kind, the existence of spinal meningitis may not be suspected during life; it may only be discovered post mortem.

The degree of pyrexia which is developed in cases of spinal meningitis is variable. In some, and especially in the purulent

cases, the temperature is high even during the earlier stages of the case. In others, as in many of the tubercular cases, it is only slight, at all events during the earlier period of the disease. In cases of this kind, the length of time which the pyrexia continues is usually longer than in most cases of myelitis. We have seen that in poliomyelitis anterior acuta, the febrile disturbance, although often very marked at the commencement of the attack, soon subsides. The same statement applies to many cases of transverse myelitis. In many cases of spinal meningitis, the pyrexia develops more gradually and persists for a greater length of time. In some cases, and especially in those cases in which the membranes covering the medulla oblongata and the upper part of the cervical cord are affected, the temperature rapidly rises and hyperpyrexia is developed before death.

Irritative symptoms.—In the early stages of spinal meningitis, irritative symptoms are usually prominent. In this respect and in the long duration of the irritative stage there is a marked contrast between spinal meningitis and myelitis. The more important irritative symptoms are:—

(1) *Pain in the back.*—As I have already pointed out, this symptom seems to be chiefly due to the irritation of the sensory nerves in the inflamed membranes. When the meningitis is general, the pain may be felt over the whole of the back; but in the majority of cases it is limited to special points, for, at the commencement of the case at all events, the inflammation is usually more marked at certain parts than at others.

The back pain is usually increased by movement, by any external irritation, by percussion of the spinal column or by the application of a hot sponge to the back. We do not nowadays attach much importance to the hot-sponge test. It is merely a test of altered sensibility (hyperæsthesia) of the skin. But since hyperæsthesia is often present in meningitis and usually absent in myelitis, the hot-sponge test (granting that the hyperæsthesia is due to organic disease of the spinal cord or its membranes) is of some importance for differential diagnosis.

(2) *Hyperæsthesia of the skin and shooting (neuralgic) pains* in the areas of distribution of the posterior nerve-roots which happen to be irritated.

(3) *Muscular cramps and spasms* in the muscles supplied by motor nerve roots which are irritated. In consequence of the

muscular tension and rigidity (and partly no doubt in consequence of the fact that movements of the spinal column excite and aggravate the pain) the spine is often kept rigid and stiff. When the inflammation involves the membranes in the upper part of the spinal canal and at the posterior part of the base of the brain, the head is apt to be retracted; this is a characteristic feature in many cases of tubercular cerebral meningitis.

(4) *Increased reflex irritability*.—This is usually a marked feature in the earlier stages of spinal meningitis. Any external irritation is apt to aggravate the muscular spasm and to increase the rigidity and stiffness of the back. In consequence of this increased reflex excitability, spasmodic contraction of the sphincters of the bladder and rectum and resulting retention of urine and obstinate constipation may be produced. Retention of urine and constipation are in some cases produced in another way. In consequence of the pain and spasm which muscular movements cause, patients affected with spinal meningitis instinctively keep as quiet as possible; they avoid emptying the bladder and rectum; under these circumstances, the bladder may become over-distended and the detrusor muscle may become paralysed.

When the respiratory muscles are the seat of spasm, difficulty in breathing may be experienced.

As the meningitis progresses, these irritative symptoms become more prominent; the head may be so much retracted and the spasm of the neck muscles so great, that difficulty of breathing, with stridor and even pain and difficulty in swallowing may be produced.

In some cases, the back becomes not only rigid, but so markedly arched that opisthotonos is produced.

The irritative symptoms are, as I have already remarked, chiefly characteristic of the early stages of the disease.

Paralytic symptoms.—After a little time (usually some days), the irritative symptoms are associated with, or replaced by, paralytic symptoms. The more important paralytic symptoms are:—

1. *Anæsthesia* in the areas of distribution of the posterior nerve roots which are implicated.

2. *Paralysis* of the muscles supplied by anterior nerve roots the function of which is interrupted by the inflammatory process.

3. *Diminution or abolition of the reflexes.*

4. *Paralysis of the bladder and rectum.*

In some cases, the paralytic symptoms are, of course, partly due to implication of the tissues of the cord itself (myelitis).

Provided that the patient survives a sufficient length of time, bedsores may be developed during this, the second or paralytic, stage of the disease.

It is important to remember that there is no sharp line of demarcation between the irritative and paralytic periods of the disease. The two run insensibly one into the other. Even in the earlier stages of the case when the irritative symptoms are prominent, some anæsthesia and some muscular paralysis may be present.

As the disease advances, considerable emaciation is usually produced. In the later stages of the case, the condition of the patient may be pitiable in the extreme.

The condition of the paralysed muscles of course varies with the stage of the disease and the severity of the attack. In the earlier stages, the emaciation and muscular wasting are general; but in the later stages of long-continued cases marked and localised atrophy may be developed in some of the paralysed muscles. Those muscles which are supplied by anterior nerve roots which are seriously damaged will of course become atrophied. In the terminal stages of long-continued cases, contractures may be developed; the thighs, for example, may be flexed on the abdomen, and the legs on the thighs.

The mental condition of patients affected with spinal meningitis is not usually disturbed; an exception to this statement, of course, occurs in those cases in which the cerebral meninges are implicated.

Duration and termination.—The duration of cases of acute spinal leptomeningitis is variable. In the more severe cases, and especially in the purulent form, the disease may prove fatal within the course of two or three days; but in most cases the duration is considerably longer than this—a week, ten days, a fortnight, three weeks or even longer. In some cases, the acute meningitis gradually subsides and passes into a chronic condition.

Septic and tubercular cases are almost always fatal. In the

epidemic form (epidemic cerebro-spinal meningitis), recovery is by no means uncommon. I shall afterwards refer in more detail to this form of the disease, and I shall then point out that cases of epidemic cerebro-spinal meningitis present great differences in their severity and course.

The more severe cases of acute sero-fibrinous (simple) leptomeningitis not unfrequently terminate in death. In the less severe cases, especially in those cases in which the meningitis seems to be idiopathic or rheumatic (the result of cold) or due to traumatic injury, recovery not unfrequently takes place. In many of these cases, the acute symptoms gradually subside and the case becomes chronic. In the cases which recover or become chronic, paralyses and contractures may remain. The extent and distribution of the paralysis of course depend upon the number of nerve roots which are implicated, the extent to which they are damaged and the nature and severity of the complications (inflammatory changes) which are developed in the spinal cord itself.

In septic cases, and in some cases of epidemic cerebro-spinal meningitis, the intensity of the inflammation causes death within two or three days from the onset of the attack. In other cases, the fatal termination is due to the development of cerebral complications (coma, increased intracranial pressure, paralysis of the respiratory centre, etc.) As I have already pointed out, hyperpyrexia is not unfrequently developed just before death. In some cases, the immediate cause of death seems to be spasm or paralysis of the respiratory muscles. In cases which run a prolonged course, in many of the cases for example in which the acute symptoms subside and the inflammation becomes subacute or chronic, the patient ultimately dies from exhaustion, from bed-sores or from the development of urinary complications.

Diagnosis.—In well-marked and typical cases of acute spinal leptomeningitis the diagnosis is not difficult. We have seen that acute spinal leptomeningitis is a febrile disease, which is attended with well-marked nervous symptoms indicative of derangement of the functions of the spinal cord and of the nerve roots. The disease is characterised by :—(1) an irritative stage, the chief symptoms of which are pain in the back (increased by movement and percussion), rigidity of the spine, hyperæsthesia,

shooting pains in the limbs, cramps and spasms in the muscles of the limbs and trunk, constipation, and in some cases retention of urine: and (2) by a paralytic stage, the chief symptoms of which are anæsthesia, motor paralysis, diminished reflex excitability and paralysis of the bladder and rectum. The irritative and paralytic stages run insensibly one into the other. When such a combination of these symptoms is present the diagnosis is clear enough.

The diseases which are most apt to be mistaken for acute spinal leptomeningitis are tetanus and myelitis.

The differential diagnosis of acute spinal meningitis and of tetanus.—Tetanus is perhaps more likely to be confounded with acute generalised spinal meningitis than any other condition; but the differential diagnosis is not difficult. In cases of traumatic tetanus, there is usually a history of a wound or injury, often of a fall on earth, for the tetanus organism seems to have its resting-place in the soil, or the presence of a wound. In tetanus, there are no sensory symptoms, for the motor side of the nerve apparatus is alone affected. In tetanus, trismus is usually a very marked and characteristic feature; in cases of cerebro-spinal meningitis, trismus is sometimes developed, but is very rarely indeed (if ever) due to an inflammation confined to the spinal membranes. Opisthotonos, which is such a characteristic feature of tetanus, is only exceptionally present in cases of spinal meningitis. The paroxysmal exacerbation of the muscular spasms, which are often attended with extreme cyanosis and which are not unfrequently fatal, are characteristic symptoms in well-pronounced tetanus, but are rarely, if ever, present in acute spinal leptomeningitis. In tetanus, the reflex irritability is enormously exaggerated, and any external or peripheral irritation is apt to excite a violent attack of spasms. In a minor degree the same increased reflex excitability is observed in some cases of spinal meningitis, but the paroxysmal exacerbations of the spasms are, comparatively speaking, very slight. In tetanus, the course of the disease is, speaking generally, more rapid than that of spinal meningitis. In tetanus, paralytic symptoms are seldom, if ever, developed.

From these statements, you will readily understand that there is usually no difficulty in distinguishing the two affections.

The differential diagnosis of spinal meningitis and myelitis.—In speaking of myelitis, I have already pointed out the chief points of distinction between spinal meningitis and myelitis. The marked character of the irritative symptoms, the long duration of the irritative stage, and the absence (at all events until the later stages of the disease) of paralysis of the sphincters and of bed-sores are the most important points.

The differential diagnosis of the different forms of leptomeningitis.—The final step in the diagnosis of spinal meningitis is to determine the variety or form of the disease which is present—whether the meningitis is of the simple or sero-fibrinous kind; whether it is purulent, septic or of the epidemic variety; or whether it is tubercular. In many cases it is extremely difficult or impossible to decide this point, which is nevertheless of considerable importance, for the prognosis undoubtedly differs in these three great groups.

Acute simple leptomeningitis with a sero-fibrinous exudation is, in some cases, the result of exposure to cold and wet, and occasionally, but rarely, of traumatic injury; it may follow influenza. It may originate without any definite exciting cause or associated pathological condition—a negative fact which is of some diagnostic importance. But it must be remembered that this form of acute spinal meningitis is rare. In most cases, the onset is gradual and the course more or less prolonged—as a rule much less rapid than in the purulent cases. The temperature at the commencement is rarely very high. The irritative symptoms are usually marked. The inflammation is usually confined to the spinal meninges, and cerebral symptoms are therefore absent. It must however be remembered that in many of the milder cases of the epidemic cerebro-spinal form the inflammation is probably sero-fibrinous (not purulent); the complete recovery seems to show this.

In the *purulent form*, an associated septic condition, such as pyæmia, ulcerative endocarditis or pneumonia is often present; in many of the grave cases of epidemic cerebro-spinal meningitis, the exudation also consists of pus. In many of the purulent cases, the onset is abrupt and often attended with a rigor. In these cases, the temperature is apt to run up rapidly, and cerebral symptoms are often present, for the cerebral meninges are in many cases also affected. Further, in the purulent cases,

irritative symptoms are often less marked, or at all events less apparent than in the other varieties of the disease; they seem in some cases to be overshadowed by the severity of the general constitutional disturbance. It is important to remember that a purulent inflammation does not, as a rule, seem to produce so much irritation of the sensory nerves in the inflamed membranes as a sero-fibrinous inflammation does. I have already emphasised this point in describing pericarditis and peritonitis. I have several times found a purulent peritonitis after death in cases in which there was absolutely no pain, and, I may also add, very little pyrexia during life.

In the *tubercular form* of spinal meningitis, the patient is much more frequently a child than an adult. In these cases, symptoms indicative of cerebral meningitis are generally conspicuous; in fact, as I have already pointed out, the spinal symptoms are in many cases so entirely overshadowed by the cerebral symptoms that the presence of a tubercular inflammation of the spinal membranes is not suspected during life. In the exceptional cases of tubercular spinal meningitis in which the cerebral symptoms are inconspicuous or only developed in the later stages of the case, the course is usually protracted and the symptoms less severe than in the purulent form. The temperature is rarely high, in the early stages at least. In tubercular cases, the general constitutional condition of the individual may afford a clue to the nature of the spinal inflammation. In some cases, there are associated tubercular lesions in other organs, the lungs for example; in others, the tubercular inflammation of the spinal membranes is secondary to a tubercular tumour in the substance of the cord (it is usually situated in the lumbar enlargement). In some, the family history or the general physique and constitutional condition of the patient are suggestive of the tubercular nature of the inflammation.

LECTURE XXXII

ACUTE SPINAL LEPTOMENINGITIS (*Continued*)

IN the last lecture, Gentlemen, we considered the diagnosis of acute leptomeningitis. Let us now turn to the prognosis and treatment.

Prognosis.—Acute general spinal meningitis is always a very grave disease. In the great majority of cases in which the inflammation is severe, the result is fatal.

In trying to judge of the severity of the attack in any particular case, we have to take into account:—

(1) *The extent and severity of the inflammation.* High fever at the commencement indicates a severe attack. When the irritative symptoms involve both the upper and the lower limbs and the trunk, when, in other words, the clinical symptoms show that the inflammation is generalised all through the spinal canal, the case is, other things being equal, more severe than in those cases in which the inflammation is localised and limited in extent. The presence of head symptoms, which show that the cerebral meninges are implicated, is of grave significance. When the respiratory muscles are involved, there is always great danger.

(2) *The type of the inflammation.* As I have already said, it is often a difficult or impossible matter to come to any definite conclusion on this point during life. A simple sero-fibrinous inflammation is, of course, less serious than a purulent or tubercular inflammation. I do not mean to say that a tubercular inflammation of the spinal membranes is necessarily fatal, for there is good reason to suppose that tubercular cerebral meningitis is sometimes recovered from. But under our present methods of treatment recovery is so rare that in cases of tubercular spinal meningitis, which can be definitely recognised and

diagnosed during life, the prognosis is practically speaking hopeless. A general purulent inflammation of the spinal membranes is probably always fatal. If then the spinal meningitis seems to be due to a septic cause, or if there is reason to suppose that the inflammation is purulent, a fatal issue must, in the present position of our knowledge and methods of treatment, be expected. The prognosis of the epidemic form of cerebro-spinal meningitis depends upon the severity of the attack; the severe cases usually prove fatal; the slight cases often recover or pass into a chronic condition. This point will be considered in more detail when I come to speak of this variety of the disease.

(3) *The rapidity with which the symptoms are developed and with which the disease is progressing, the presence or absence of grave complications, and the effects of treatment.* The early development of paralytic symptoms and the presence, in the later stages and in cases in which the inflammation becomes chronic, of paralysis of the sphincters and of bed-sores, are of grave significance.

(4) *The age and previous state of health of the patient.* Though these points are less important in spinal meningitis than in many other diseases, they have to be taken into account.

Treatment.—The same measures of treatment which have been recommended in the earlier stages of acute myelitis, with the object of cutting short and arresting the inflammatory process, should be employed. I need not go into any great detail. It will be sufficient to say that dry cupping over the spine and the application of hot fomentations or an ice-bag to the spinal column are probably the most important local means of treatment. The ice-bag is only to be recommended in those cases in which it can be continuously kept in contact with the spinal column (a matter in some cases of considerable difficulty) and in which it is comfortably borne by the patient. In those cases in which the ice-bag seems to aggravate the patient's sufferings, hot fomentations are preferable.

A brisk purge should be administered at the commencement of the attack, and those internal remedies, which experience has shown to be useful in the special form of inflammation which happens to be present, should be employed.

Tartar emetic, mercury, iodide of potassium, salicylate of soda and salol are perhaps the most useful drugs in those cases in which the inflammation appears to be of the sero-fibrinous type and in which it seems to be idiopathic or rheumatic in origin (due to exposure to cold and wet).

Quinine, boracic acid, salol, and the tincture of the perchloride of iron are probably the most important and useful remedies in septic cases and in those cases in which the inflammation is of the purulent type.

Iodide of potassium is perhaps the only internal drug which, so far as our present therapeutic knowledge enables us to judge, is likely to be beneficial in the tubercular cases. In this form of the disease, iodoform ointment should be rubbed over the back and scalp. We shall afterwards see that there is some reason to believe that the frequent inunction of iodoform ointment into the scalp is beneficial in some cases of tubercular cerebral meningitis.

In addition to the local and general measures which have just been recommended with the object of restraining and subduing the inflammatory process, special means must be taken to relieve the pains and spasms which are so distressing to the patient. The spasms are best relieved by bromide of potassium, chloral hydrate or a combination of these drugs; and the pain by morphia, phenacetin and antipyrin.

It is unnecessary to say that the patient must be kept at absolute rest; you do not require to tell him to keep quiet, he instinctively keeps as quiet as possible, for he knows that the slightest movement or exertion is apt to increase the pain and aggravate the spasm. But it may be, and usually is, essential to impress upon the nurse the importance of avoiding all movement and everything which is likely to cause peripheral irritation. The patient should be allowed to lie in the position which is most agreeable to himself. Here, as in other cases, it is rarely wise to interfere with the indication which Nature gives us. The patient usually lies on his back with his legs drawn up, and the head perhaps retracted.

The condition of the bladder requires to be carefully attended to. We have seen that at the beginning of the disease retention of urine may be due to spasm of the sphincter. The remedies most calculated to relieve this spasm are morphia suppositories

and warm fomentations locally; and chloral hydrate, bromide of potassium and morphia given by the mouth. It is only when these measures do not succeed that recourse should be had to the catheter.

The diet must of course be liquid (milk, strong beef extracts, white of egg, soups, etc.).

In septic cases, and in the later stages of the other forms of the disease, stimulants may be necessary.

In those cases in which the acute stage subsides and the condition becomes chronic, counter-irritation of the spine (blisters, painting with iodine, in some cases the application of the actual cautery) and the internal administration of iodide of potassium, the syrup of the iodide of iron, together with good feeding, tonics (quinine and cod-liver oil) are the most important means of treatment.

When there is reason to suppose that the inflammation has subsided, local treatment must of course be employed with the object of preventing and removing the contractures and restoring the nutritive condition of the atrophied and paralysed muscles. Massage, electricity, nux vomica, or strychnine are useful remedies at this period of the disease; but it is essential to remember that they are apt to be harmful rather than beneficial if commenced at a too early period of the case.

CHRONIC SPINAL LEPTOMENINGITIS

The next affection to which I wish to direct your attention is chronic spinal leptomeningitis. This is a much less definite and distinct disease than the acute variety. In many cases in which the soft membranes of the spinal cord are affected with chronic inflammation, the symptoms are very obscure and the diagnosis attended with great difficulty.

Etiology.—Chronic leptomeningitis may follow an acute or sub-acute inflammation of the soft membranes of the spinal cord or it may be chronic from the first. In the latter group of cases, the inflammation of the soft membranes is usually secondary to a primary lesion in the spinal cord itself or to Pott's disease of the vertebræ and pachymeningitis externa.

In the rare cases in which the inflammation is primary (i.e. originates independently of any definite intra- or extra-medullary lesion), exposure to cold, syphilis, alcoholic excess and traumatic injury are probably the chief causes of the condition. Possibly these cases are more common than many of us are disposed to think, but the difficulty in recognising them with certainty during life makes it impossible to speak more definitely. All I can say is, that so far as my experience enables me to judge, chronic inflammation of the soft membranes of the spinal cord, the result of the causes which have just been enumerated (syphilis and severe traumatic injuries excepted) is an exceedingly rare condition. Occasionally a deposit of tubercles in the spinal membranes seems to produce a chronic rather than an acute or sub-acute inflammation of the soft membranes; but this is certainly rare.

It is probable, then, that all the conditions which produce acute leptomeningitis may, when acting in a less intense degree, cause chronic inflammation of the soft membranes.

Chronic leptomeningitis is usually more limited and localised than acute leptomeningitis. An exception to this statement may, of course, occur in those cases in which the chronic inflammation of the soft membranes is secondary to the acute form of the disease; but even in these cases, the more marked inflammatory changes are usually localised to special parts, the reason, of course, being that at places the acute inflammatory changes have subsided and entirely disappeared without leaving any definite and distinct alterations (adhesions or thickenings) behind them. The chronic changes which remain are only apparent in those positions in which the acute inflammation was most intense.

Morbid Anatomy.—In cases of chronic leptomeningitis, the spinal fluid may be increased in amount; it is sometimes turbid, but more often clear. The most characteristic appearances are thickenings and opacities of the soft membranes and the presence of adhesions which bind the membranes to one another and to the surface of the spinal cord. The nerve roots may be surrounded and ensheathed by adhesions and masses of cicatricial tissue. In some cases they are compressed and atrophied; but these changes in the nerve roots are usually less marked

than in pachymeningitis. I am referring more particularly to those cases of chronic leptomeningitis in which the inflammation arises independently of a pre-existing acute attack. In those cases in which the chronic leptomeningitis is merely an after-result of an acute attack, the nerve roots may of course be extensively and severely damaged. In the syphilitic cases, too, in which a meningitis develops around a localised gumma, the nerve roots which are implicated and infiltrated by the gummatous deposit may be severely damaged. In cases of this kind, the membranes around the gumma are usually adherent to the surface of the cord, and the tissues of the cord itself are involved in a greater or a less degree. In these cases, the gummatous myelitis is much more important than the gummatous meningitis.

It would appear that in some cases a chronic inflammation of the membranes on the surface of the cord is the starting-point of a slow sclerotic lesion in the cord itself. We have seen that locomotor ataxia is perhaps in some cases produced in this way.

Pathological Physiology.—The remarks which I have already made with regard to the pathological physiology of acute leptomeningitis apply to the chronic variety of the disease. The symptoms in chronic leptomeningitis are partly due to irritation of the sensory nerves in the inflamed membranes, partly to implication of the anterior and posterior nerve roots, and partly to implication of the cord itself.

Since the irritation is less intense and more localised than in the acute form, you can easily enough understand that the pain in the back is usually less severe and more limited in its distribution. In many cases of chronic leptomeningitis the nerve roots are only involved in a slight degree; the root-symptoms are rarely so marked and never so widely distributed over the surface of the body as in the acute form of the disease. Further, the symptoms indicative of reflex and motor irritation (muscular cramps and spasms) are much less prominent than in the acute variety. Usually, too, when the case first comes under observation the irritative and destructive symptoms are more or less mixed up. It is seldom in the chronic form that a definite and distinct stage of irritation precedes a definite and distinct stage of

paralysis. Again, in those cases in which a chronic inflammation of the soft membranes is secondary to an intra- or extra-medullary lesion, the clinical picture is a complicated one. In these cases the symptoms due to the primary disease are usually far more conspicuous than those which result from the chronic leptomeningitis.

Clinical History.—In those cases in which chronic leptomeningitis follows acute leptomeningitis, there is no sharp and abrupt line of transition between the two conditions. In those cases in which the leptomeningitis is secondary to pachymeningitis externa and Pott's disease of the vertebræ, symptoms indicative of the primary lesion are usually conspicuous. In those cases in which the chronic leptomeningitis is associated with myelitis or any other intra-medullary lesion, the symptoms due to the cord lesion usually occupy the chief place in the clinical picture.

The symptoms which are due to chronic leptomeningitis itself are in many cases indefinite and obscure. In well-marked cases, the onset is usually gradual and unattended with fever. Pain in the back, increased by movement, by percussion and by the hot sponge test, is a characteristic symptom; but it is usually more localised and less intense than the pain of acute leptomeningitis. Further—and this is a very important point from a diagnostic point of view—the pain in the back is not attended with any local swelling, prominence of the vertebral spines or spinal curvature. There may be some rigidity of the spinal column, but it is usually much less marked than in the acute form of the disease. Shooting pains and hyperæsthesia in localised parts of the body (in the sensory areas of the posterior nerve roots which happen to be irritated) are present in some cases; but they are usually less marked than in acute leptomeningitis and in chronic pachymeningitis. In the great majority of cases, the symptoms indicative of sensory irritation are associated with more or less anæsthesia. In some cases, the remarkable condition termed *anæsthesia dolorosa* (in which tactile and painful impressions are not perceived, but in which severe pains are complained of and spontaneously developed in the anæsthetic parts) is present over a limited area or areas of the body.

Anæsthesia dolorosa is highly suggestive of pressure either on the posterior nerve roots or the sensory peripheral nerves. Though occasionally perhaps met with in chronic leptomeningitis, it is much more common in pachymeningitis (externa and interna hypertrophica) and in cases in which the posterior nerve roots or peripheral nerves are pressed upon or invaded by a new growth—a tumour, for example, in the lower part of the spinal canal which presses upon the cauda equina, or a tumour in the pelvis which involves the nerves going to the lower limbs. You will observe that in both of these cases motor paralysis, which is usually paraplegic in distribution, is necessarily associated with the peculiar form of anæsthesia to which I am referring. A tumour which presses upon and involves the nerve strands of the cauda equina will necessarily produce both motor and sensory paralysis and will involve the nerve fibres going to both lower extremities. A tumour in the pelvis may of course involve the nerves going to one limb only, but often involves the nerves proceeding to both lower limbs. These facts explain the frequent association of paraplegia with this peculiar form of anæsthesia dolorosa. ‘Painful paraplegia’ is, in fact, almost synonymous with paraplegia due to pressure on peripheral nerves or nerve roots, and is very frequently due to disease of the spinal bones, often to malignant disease of the vertebræ or malignant growths in the pelvis.

You will find these general propositions useful in dealing with cases of paraplegia in which this peculiar form of anæsthesia is present. Before leaving the subject, let me briefly refer to the manner in which anæsthesia dolorosa is produced. Under normal circumstances, a tactile impression on the surface of the skin excites a nerve impulse which passes along the peripheral nerve, through the spinal cord, medulla oblongata, etc., until it reaches the sensory perceptive centre in the cerebrum. The impression which is generated in the centre is referred by the sensorium to the area of skin which was touched or irritated. Now, a lesion which interrupts the conduction through the sensory peripheral nerve will of course produce anæsthesia in the area of distribution of this sensory nerve, for impressions passing from the periphery will be blocked at the point of lesion. But suppose that in addition to the interruption of function, irritation is produced at the upper level

of the lesion. In consequence of this irritation of the sensory nerve fibres at the upper level of the lesion, nerve impulses will be generated and carried to the sensory perceptive centre in the cerebrum; the conscious impressions which are thereby generated will be painful, and the pain will be referred, not to the seat of irritation (say the cauda equina), but in accordance with the law of 'eccentric projection,' to that portion of the surface from which the nerve fibres which are irritated are in the normal condition of things in the habit of conducting impulses and to which the sensorium is under normal circumstances in the habit of referring the sensations which are generated by those impulses. It is in consequence of this law of 'eccentric projection' that patients after amputation often feel pain in the toes, which may have been dead and buried years before, and that in some cases they are able to describe the exact position in which the fingers or the toes of the amputated limb happen to be placed. It is a remarkable fact that by faradising the nerves in the stump conscious sensations of definite movement, in the limb which is no longer present, can in some cases be aroused in the sensorium; and that these conscious impressions correspond exactly to the conscious impressions which would have attended the actual muscular movements which faradic stimulation of the nerve would have produced in the muscles of the limb which has been removed. In other words, irritation of the individual nerves in the stump is attended with exactly the same conscious impressions in the sensorium which would have resulted if the muscular movements had actually taken place. In consequence of these conscious impressions the patient says that the limb (which is no longer present) is placed in certain definite positions, which exactly correspond to the position which stimulation of the nerve would have produced if the limb were still present.

Remember, then, that although the condition which we term *anæsthesia dolorosa* is present in some cases of chronic leptomeningitis, it is much more frequently the result of pachymeningitis or of the pressure of a tumour on the nerve roots or the peripheral nerve trunks.

In addition to the sensory symptoms which I have just described, motor symptoms, both of an irritative and a paralytic kind, are usually present in cases of chronic leptomeningitis.

These irritative motor symptoms are usually less prominent than in acute leptomeningitis; some muscular stiffness and rigidity is not uncommon, but the more definite cramps and spasms which are so frequent in acute leptomeningitis are rarely observed. In cases of chronic leptomeningitis, some muscular weakness, it may be some definite paralysis, is usually present when the patient first comes under the observation of the physician. I repeat that in many cases of chronic leptomeningitis the root-symptoms are by no means prominent.

There is an exception in the syphilitic variety of the disease, but this is perhaps hardly a true exception, for, as I have already pointed out, the syphilitic form of chronic leptomeningitis is in many cases associated with, and the result of, a gumma which for practical purposes may be regarded as a tumour surrounded by inflammation. Further, in many cases of chronic leptomeningitis paralytic symptoms are the result of lesions in the cord itself; but to those cases I have already referred.

As the disease progresses, the anæsthesia and the paralysis become more marked. In the earlier stages, there may be little or no alteration in the condition of the muscles, but in the advanced stages well-marked atrophy, limited to the area of distribution of the anterior nerve roots which happen to be involved, may be developed.

The deep reflexes are in some cases diminished or abolished; in others exaggerated.

The bladder and rectum are rarely affected in any marked degree, unless the inflammation happens to involve the nerve strands of the cauda equina or the lower lumbar and sacral regions of the cord.

Summary of the symptoms characteristic of chronic spinal leptomeningitis.—To sum up, then, the symptoms which may be present in cases of chronic leptomeningitis are:—

(1) Pain in the back, increased by movement and percussion and often localised to individual parts of the spinal column.

(2) Root-symptoms, such as shooting pains, anæsthesia, hyperæsthesia, gradually progressive weakness in individual muscles, abolition of some of the reflexes, increased tonicity (rigidity), or more frequently atrophy in localised parts of the body. I must ask you to note that some of the root-symptoms which have just been enumerated are often absent, and that in

many cases of chronic leptomeningitis the root-symptoms are by no means marked.

(3) Symptoms of myelitis.

(4) In some cases, symptoms due to disease of the bones or other primary lesion to which the leptomeningitis is secondary.

The condition of the general health is variable; it is impossible to make any general statement which is applicable to all cases. In those cases in which the inflammation follows a severe attack of acute leptomeningitis, emaciation and feebleness are often conspicuous; in many of these cases, the patient dies after some weeks or months, worn out by exhaustion, by bed-sores or by kidney or bladder complications. But in other cases the general state of nutrition is fairly good.

Diagnosis.—The diagnosis is often attended with great difficulty. In those cases in which the leptomeningitis is the result of a syphilitic gumma or other well-marked form of extra-medullary lesion, the nature of the case is usually clear enough.

In cases, too, in which the leptomeningitis is combined with myelitis, the diagnosis is not as a rule difficult. What I mean to say is, that in cases of this kind there is usually no great difficulty in deciding that the symptoms are due to organic disease. But it is often a very difficult matter to determine whether a pachymeningitis is complicated with leptomeningitis, and in cases of meningo-myelitis to form a correct opinion as to the extent and severity of the leptomeningitis which is present.

The cases of chronic leptomeningitis in which the diagnosis is most difficult are the least severe cases and those in which the leptomeningitis is unassociated with myelitis or any localised extra-medullary lesion.

The differential diagnosis of chronic leptomeningitis and hysteria.—The distinction is sometimes very difficult. The points to which attention should be chiefly directed are:—

The age and sex of the patient.—In hysteria, the patient is usually a woman, and generally a young woman; whereas in the cases of chronic leptomeningitis to which I am now referring the patient is more frequently a male than a female, and generally a middle-aged male.

The exact character of the symptoms.—In hysteria the pain in

the back is less constant, subject to greater variations in its extent and distribution, and usually associated with a greater degree of hyperæsthesia of the skin; the slightest touch on the back in some hysterical cases gives rise to complaints of great pain and suffering. In some of these cases, firm pressure, especially when the patient's attention is directed to something else, is well borne. Muscular movement is less apt to aggravate the back-pain of hysteria, but it is important to note that in many cases of traumatic neurasthenia following railway accidents and injuries (in which there is absolutely no evidence of spinal meningitis) the back is kept stiff and the back-pain is increased by movement. Then again, although in hysteria shooting neuralgic pains may be complained of, and localised or more widely diffused areas of hyperæsthesia or anæsthesia may be detected in various parts of the body, these sensory symptoms are rarely limited and localised to the areas of distribution of individual posterior nerve roots. The same statement applies to the motor symptoms. In hysteria, the muscular weakness and paralysis (if distinct paralysis is present) are rarely localised and limited to the areas of distribution of definite motor nerve roots. In short, in hysteria the subjective symptoms are usually out of all proportion to the objective symptoms which can be demonstrated; whereas, in organic disease there is usually more or less proportion between the objective and the subjective symptoms. This is a very important point.

Then again, in hysteria localised muscular atrophy is rarely if ever present except as an associated condition, i.e. the result of an associated lesion such as a localised neuritis.

The presence of associated symptoms indicative of hysteria.—Further, in hysterical cases other symptoms and signs of hysteria are usually present. The fact that they exist is not of course conclusive; for, as I have again and again told you, hysterical symptoms are not infrequently associated with organic disease. But the presence of well-marked hysterical symptoms in the absence of any definite symptoms and signs indicative of organic disease is of great diagnostic significance. Again, in hysteria the symptoms are apt to fluctuate from time to time, though this is by no means always the case. The presence or absence of a cause of hysteria, such as uterine or ovarian disease, must also be taken into account.

In doubtful cases, an experienced and shrewd observer can generally come to a correct conclusion, the opinion being in many cases largely based upon the general impression which his clinical experience enables him to draw from the whole facts of the case.

The differential diagnosis of chronic leptomeningitis and locomotor ataxia.—In its earlier stages, locomotor ataxia may in some cases perhaps be confounded with chronic leptomeningitis. This is not to be wondered at when it is remembered that in some cases of locomotor ataxia the soft membranes over the posterior columns of the cord are actually inflamed. But even in those cases of locomotor ataxia in which the leptomeningitis is most marked there is generally no difficulty in coming to a correct conclusion as to the nature of the case. In locomotor ataxia, the lightning pains are much more marked than in chronic leptomeningitis, and they are usually confined to the lower extremities and the lower parts of the trunk. Further, in locomotor ataxia the knee-jerks are almost invariably absent, the Argyll Robertson condition of the pupil is often present, and there is often some derangement of the vesical, rectal, or sexual reflexes. The presence of the characteristic inco-ordination is of course conclusive. Again, in locomotor ataxia, pain in the back is seldom so marked as in chronic leptomeningitis.

Prognosis.—From what I have just said as to the difficulty of definite diagnosis, you will readily understand that the prognosis is in many cases a matter of difficulty and uncertainty. In the absence of a definite diagnosis, the prognosis must always be uncertain, for a correct diagnosis is the only sure basis on which the prognosis can be founded, together of course with knowledge and experience as to the course which any particular disease or pathological process is likely to pursue, and the all-important considerations which belong to the personal equation and which guide the opinion in individual cases.

A chronic inflammation of the soft membranes of the spinal cord is never a trivial matter. Once established, the inflammation is apt to pursue a progressive course. Further, it may be associated with a cord lesion or with a lesion of the bones which, when the patient first comes under observation, is unattended

with any very definite symptoms, or it may be the starting-point of a cord lesion.

In individual cases, the cause of the meningitis, its extent and severity, the effects of treatment, whether it seems to be progressive or not, the presence or absence of complications and associated lesions, the general condition of the patient and the state of his previous health have to be taken into account.

Treatment.—The general health must be kept in the highest possible state of efficiency, and the local and general measures which are advisable in acute leptomeningitis after the acute stage has subsided should be employed. Counter-irritation, the application of blisters or the actual cautery over the affected part of the spine; the internal administration of iodide of potassium, mercury, syrup of the iodide of iron; general tonics; and in those cases in which there are localised contractures or paralyses, massage and other suitable means of local treatment are the measures on which we chiefly rely.

LECTURE XXXIII

PACHYMENINGITIS SPINALIS

Introductory remarks.—To-day, Gentlemen, I propose to direct your attention to inflammation of the spinal dura mater (pachymeningitis spinalis). I have already told you that an inflammation which commences in, or is confined to, the spinal dura mater is almost always chronic in its course, usually localised in extent and generally limited to the outer surface of the dura. Further, we have seen that inflammation of the inner surface of the dura is frequently associated with acute leptomeningitis; but that the inflammatory process comparatively rarely makes its way through the thick, tough, resisting dura mater.

The external and internal forms of inflammation of the spinal dura mater are termed pachymeningitis spinalis *externa* and *interna* respectively.

ACUTE GENERALISED SUPPURATIVE PACHY- MENINGITIS EXTERNA

Etiology and Pathology.—Pachymeningitis spinalis externa is very rarely acute. In the vast majority of cases it is a chronic affection which is secondary to a lesion of the bones, such as Pott's disease of the vertebræ—its most common cause. Cases are, however, occasionally but very rarely met with in which an acute suppurative inflammation involving the connective tissue on the outer surface of the dura mater (i.e. between the dura and the bone) extends over an extensive area, it may even be from the top to the bottom of the spinal canal. A suppurative inflammation of this kind may be due to the bursting of an abscess or a deep bed-sore, for example, into the spinal canal, or may be part and parcel of a general septic state, the septic irri-

tant being perhaps carried by the blood-vessels to the venous plexus which is present in the connective tissue between the dura and the bone.

One might perhaps expect that an acute and diffused inflammation on the outer surface of the dura would occasionally result from a localised purulent or caseous collection connected with Pott's disease of the vertebræ—a very common condition ; but this rarely if ever happens, the reason being that the pus is hemmed in and localised by inflammatory adhesions. No case of acute generalised suppurative inflammation on the outer surface of the dura mater has come under my observation.

Clinical History.—Acute suppurative inflammation of the dura, if generalised, is attended with marked fever and constitutional disturbance ; rigors are often present. The local symptoms and signs are :—Pain and tenderness on pressure ; œdema of the back along the sides of the spinal column, and occasionally perhaps local abscess formations with fluctuation. Root-symptoms might of course be present from implication of the nerve roots ; and if a localised collection of pus should form between the dura and the bone the spinal cord might, of course, be pressed upon and cord symptoms might be developed. I need not go into further details which, so far as my knowledge is concerned, are entirely theoretical.

Diagnosis.—An inflammation of this description is so rare that it may be said to be one of the curiosities of medicine. I have never seen a case in which I had any reason to suppose that it was present, and even if it were present it would, I conceive, be very difficult to diagnose. The symptoms and signs suggestive of the condition would be :—Fever, great constitutional disturbance, localised pain in the back, and perhaps nerve symptoms pointing to irritation of a number of nerve roots or to pressure on the cord itself.

Prognosis.—The condition is probably always fatal.

Treatment.—So far as I know, therapeutic measures are of little or no avail. The chief indications are :—To reduce the fever and to administer remedies which are useful in other sup-

purative and septic conditions ; to relieve the pain ; to endeavour to evacuate the pus ; and to support, feed and stimulate the patient.

PACHYMENINGITIS EXTERNA CHRONICA

Let us now pass to the chronic form of pachymeningitis externa—a common and important condition.

Etiology.—In the great majority of cases of pachymeningitis externa chronica, the inflammation is localised in distribution and secondary to some pre-existing disease of the bones or other structures outside the dura mater. Pott's disease of the vertebræ is by far the most common cause. Occasionally the inflammation is secondary to tumours, especially malignant growths affecting the vertebræ, aneurisms which have eroded the vertebral column and are pressing upon the outer surface of the dura, deep bed-sores and ulcers which have made their way into the spinal canal.

Morbid Anatomy and Pathological Physiology.—The inflammation is almost always localised and limited to the immediate neighbourhood of the primary disease, whatever it may happen to be. The outer surface of the dura mater and the connective tissues between the dura and the bone are thickened and infiltrated with inflammatory products. In cases of Pott's disease the inflammatory exudation is often caseous or purulent. The nerve roots which pass through the inflammatory or caseous collection are frequently implicated—irritated, or subjected to injurious pressure. The inflammation rarely extends through the thick fibrous dura mater ; it is generally limited to the outer surface of the membrane, but this does not prevent the cord being pressed upon. Inflammatory changes are often produced in the cord tissues as the result of this pressure ; in fact, pachymeningitis externa and the primary conditions which give rise to it are by far the most common causes of pressure paraplegia, or compression myelitis as it is sometimes termed.

Clinical History.—The symptoms which are met with in cases of pachymeningitis externa are due (1) partly to the primary

disease, such as Pott's disease of the vertebræ to which the pachymeningitis is secondary; (2) partly to the inflammation of the dura mater itself; and (3) partly to involvement of the nerve roots, pressure upon the spinal cord, and the irritative, destructive and inflammatory changes which are thereby established.

The manner in which these different symptoms are arranged or grouped and the order in which they are developed in any given case are very variable.

In most cases, symptoms due to the localised bone disease, or the primary lesion whatever it may happen to be (such as:—pain in the back, increased on movement and rotation of the spinal column; stiffness and rigidity of the spine; repugnance to movement; instinctive fixation of the back; emaciation; debility and other constitutional symptoms, which attend any chronic scrofulous lesion) usually precede the nerve symptoms, or are developed simultaneously with localised nerve symptoms indicative of pressure on, and implication of, one or more of the nerve roots.

In many cases, well-marked physical signs indicative of bone disease (such as prominence of one or more of the vertebral spines, angular or lateral curvatures, occasionally localised swelling and œdema together with tenderness on pressure, and in some cases the presence of a distinct fluctuating tumour) are present when the patient comes to the physician complaining of nervous symptoms.

In other cases, root-symptoms (such as shooting pains, hyperæsthesia, anæsthesia, muscular rigidity, tension, weakness, paralysis, atrophy and abolished reflex action) are developed in localised sensitive and muscular areas, before any symptoms of bone disease are complained of or any definite indications of bone disease, such as projection of the vertebral spines, are apparent.

Symptoms due to compression of the cord and compression myelitis (the nature of which I shall presently describe) may after a time be developed. But I need hardly say that in many of the cases in which a localised pachymeningitis externa is associated with Pott's disease, there are no cord symptoms.

Diagnosis.—The diagnosis of pachymeningitis externa essentially depends upon the presence of symptoms indicative of an

extra-dural lesion involving the nerve roots, and in many cases pressing upon the spinal cord itself. It must, however, be remembered that the mere presence of root-symptoms and of cord-symptoms indicative of pressure paraplegia is not sufficient to warrant a diagnosis of pachymeningitis externa. Exactly the same symptoms may be produced by the condition termed pachymeningitis interna hypertrophica and extra-medullary tumours (i.e. tumours situated on the surface of the spinal cord). Both of these conditions are very rare; hence, localised symptoms indicative of pressure on the nerve roots and spinal cord are in the great majority of cases suggestive of bone disease and localised pachymeningitis externa. The first step in the diagnosis of pachymeningitis externa chronica is, therefore, to determine whether Pott's disease of the vertebræ or other primary lesion of the bone is present. I will consider this question of diagnosis in more detail when I come to speak of pressure myelitis.

Prognosis.—The prognosis of pachymeningitis externa essentially depends upon (a) the nature of the primary lesion (whether it is curable or not); and (b), in those cases in which cord symptoms are present, the intensity of the myelitis. The localised inflammation on the outer surface of the dura is *per se* a matter of comparatively little importance. Provided that the primary lesion can be removed or cured, the pachymeningitis externa will in all probability subside and get well. As a matter of fact, the prognosis has in many cases to be largely guided by the effects of treatment. It is often impossible when a case comes under observation to say whether a complete cure will result or not. This point will be considered in more detail in connection with compression myelitis.

Treatment.—In cases of pachymeningitis externa chronica, the objects of treatment are to remove and cure the primary lesion, to relieve the cord from injurious pressure, to promote absorption of the inflammatory products which are present on the outer surface of the dura, and to cure the myelitis if myelitis is present. Further, in those cases in which root-symptoms are prominent, the pain must be allayed by appropriate means. I need not go into details; for the treatment is in many cases

synonymous with that of compression myelitis due to Pott's disease of the vertebræ—a condition to which I now propose to direct your attention.

COMPRESSION MYELITIS

In previous lectures I have more than once referred to compression myelitis; and as it is a common and important condition, it will perhaps be well to enter into a little more detail regarding it.

Any lesion which compresses the spinal cord may not only interfere with its functional activity, but may lead to the production of myelitis. The compressing lesion is usually situated on the outer surface of the dura mater; in actual practice, Pott's disease of the vertebræ is by far the most common cause of compression myelitis. I am speaking of chronic lesions. Acute compression of the cord is most frequently, of course, due to traumatic injury. I shall refer in detail to the traumatic cases when I come to speak of concussion of the spinal cord and the lesions of the spinal cord which result from traumatic violence. In rare instances, compression myelitis is due to sarcomatous or cancerous tumours of the spinal bones, syphilitic (gummatous) disease of the vertebræ or aneurisms of the thoracic or abdominal aorta which have eroded the vertebræ and made their way, as it were, into the spinal canal.

In other cases, the compression is the result of lesions within the dura mater, a tumour springing from the nerve roots or membranes, such as a syphilitic gumma on the surface of the cord or the condition termed pachymeningitis cervicalis hypertrophica. Excluding syphilitic gummata, all of these conditions are exceedingly rare.

In the case of a syphilitic gumma on the surface of the cord, the compression of the cord is only one, and often not the most important, element in the case. In cases of this kind, the symptoms are chiefly the result of the myelitis and meningitis.

Clinical History.—In cases of compression myelitis, the clinical picture is usually made up of symptoms due to—(a) the primary disease, whatever it may happen to be; (b) to pachy-

meningitis externa or interna ; (c) to involvement of the nerve roots ; (d) to compression of the spinal cord ; and (e) to the myelitis which the pressure is apt to produce.

After the full descriptions which I have given you of pachymeningitis and myelitis, it is unnecessary to go into details. It will suffice to refer to the mode of development of the cord symptoms and to briefly emphasise some of the more important points.

It is perhaps unnecessary to say that the exact character of



FIG. 165.—*Transverse section through the spinal cord in a case of compression myelitis.*
(\times about 8 diameters.)

The cord was compressed by a tumour ; its structure is entirely degenerated, the grey matter being scarcely distinguishable from the white. Numerous large blood-vessels with thick walls are seen all through the section.

a, anterior median fissure ; *b*, right anterior horn of grey matter ; *c*, space from which a blood-vessel has fallen out.

the cord symptoms varies with (a) the seat of the lesion, (b) the severity of the pressure, and (c) the rapidity with which the pressure is produced. Slow compression, especially when it is not very great, may merely interrupt the functional activity of the spinal cord ; rapid compression, especially when it is severe, is apt to produce myelitis and destruction of the cord tissues. (See fig. 165.) The result in both cases is the production of paraplegia or paralysis below the lesion. In the slighter cases, motion is usually much more markedly impaired than sensation.

In fact, in many cases there is no impairment of sensation. In the more severe cases, the symptoms may be those of an ordinary transverse myelitis.

The distribution of the paralysis, the state of the reflexes, the condition of the muscles in regard to their nutrition and electrical reactions, and the state of the bladder and rectum, vary, of course, in different cases, just as they do in cases of transverse myelitis (see page 444).

In cases of Pott's disease of the vertebræ, the dorsal region of the cord is most frequently involved, the result being a paraplegia which is usually incomplete and which tends to assume the spastic type. In those cases of Pott's disease in which the upper cervical vertebræ are affected, all four limbs may be paralysed.

In the extremely rare cases in which an aneurism compresses the spinal cord, the dorsal region is usually implicated.

In cases of pachymeningitis interna hypertrophica, the lesion is almost always situated in the cervical region.

Tumours which cause compression myelitis, whether they are situated inside or outside the dura mater, may involve either the cervical, dorsal, or lumbar regions.

Although in the great majority of cases of Pott's disease the compression of the cord and the resulting paralysis are slowly and gradually established, this is not always the case. A few cases have come under my notice in which the paraplegia was quickly developed. The rapid development of paraplegia in cases of Pott's disease may probably be due to several different causes. The most common is perhaps the formation of an abscess or the rapid formation of inflammatory products on the outer surface of the dura, with resulting acute compression of the cord. Another cause, perhaps equally common, is the development of acute myelitis. It not infrequently happens in cases in which slow compression with no marked cord-symptoms has been going on for weeks, that acute myelitis is rapidly developed in the part of the cord which is compressed. In other cases, but they are much more rare, the rapid development of symptoms seems due to the sudden falling in, as it were, of the diseased bones with the development of acute and very severe compression of the cord. This is most likely to occur in cases in which the patient has been walking about or making a sudden effort—

in short, subjecting the diseased and weakened spinal column to pressure, the back being unsupported by a jacket or mechanical support.

In most of the cases of compression myelitis due to caries of the vertebræ, there is no difficulty in the diagnosis. In the great majority of cases the symptoms and signs of Pott's disease are well marked, and the cause of the acute compression is therefore evident. But this is not always so. Cases of Pott's disease are occasionally met with in which there is no spontaneous pain, no projection of the bones, no curvature of the spinal column, and no local tenderness on percussion over the vertebral spines. In cases of this kind, pain can almost always be elicited if the spine is rotated sufficiently freely, or if a sudden jar is sent through the spine, if, for example, the patient is lifted from the ground and allowed to come down with a thump on his heels or (in cases in which the cervical vertebræ are affected) if the top of the head is forcibly struck.

As I have already pointed out, the nature of the cord-symptoms varies in different cases. The usual condition is a paraplegia with exaggeration of the reflexes. In some cases, the paraplegia exactly resembles the spastic paraplegia due to a transverse myelitis; but in the majority of cases, the rigidity is less marked and there is more muscular wasting than in typical cases of spastic paraplegia, whether primary or secondary. In my experience, slow compression of the cord with little myelitis usually produces incomplete paraplegia with exaggeration of the deep reflexes.

In many cases of Pott's disease, the root-symptoms are less prominent than in some other conditions, such as pachymeningitis cervicalis hypertrophica and tumours situated within the dura mater, which produce compression myelitis.

In some cases of malignant disease (sarcoma or a cancer) of the vertebræ, myelitis is developed rapidly without any pre-existing evidence of the primary disease. The following are the more important particulars of a remarkable case of this kind which came under my notice a few years ago:—

Transverse myelitis due to sarcoma of the vertebræ in a child aged 18 months; rapid development of paraplegia after a twist of the back.—The patient, a big fine boy aged eighteen months, was sent to me from Yorkshire supposed to be suffering from poliomyelitis anterior acuta.

Both lower extremities were completely paralysed; the muscles were flaccid and already considerably atrophied, although the paralysis had only been in existence for a comparatively short period of time. But I had no difficulty in satisfying myself that the case was not one of ordinary poliomyelitis anterior acuta, for the bladder was paralysed and the lower extremities were completely anæsthetic. On inquiring into the history I ascertained that the child had never suffered from any illness until the paralysis developed. A sprain of the back seemed to be the exciting cause of the disease, or rather perhaps to have aggravated the sarcomatous disease of the bones which was already in existence. A gentleman friend of the family had thrown the child up into the air, as we are all in the habit of doing with our children, and caught it in his arms. As the child came down, it gave a cry; the back seemed to have been hurt; but nothing was thought of the slight injury, for the child did not go on crying. The next day it seemed perfectly well; but within a day or two it was noticed that it dragged one leg. The paralysis rapidly developed without any distinct febrile disturbance (this was another point against poliomyelitis anterior acuta); within the course of a few days both lower extremities and the bladder were completely paralysed.

The diagnosis was difficult. I felt sure that it was not a case of poliomyelitis anterior acuta; the symptoms were suggestive of transverse myelitis, but that is a very rare condition in a young child. Although the child was very young for Pott's disease, I naturally thought of that condition as the most probable cause of myelitis. But the most careful examination of the back failed to elicit any evidence of bone disease. There was no pain, no tenderness either on pressure or rotation of the spinal column, no swelling, no curvature, no projection of the vertebræ. With the exception of the momentary pain which had been felt when the back was sprained in the manner I have just described, there had been absolutely no pain either in the back or any other part. I felt so much difficulty in making up my mind as to the nature of the case that I asked Dr. John Thomson to see the patient with me. He agreed that the case was one of transverse myelitis, and that there was no evidence of bone disease.

After an interval of some weeks, when I next saw the child, the paralysis was in no way improved. I now thought that there was perhaps the slightest swelling in the mid-dorsal region. It was so slight that I did not feel sure about it; but I felt so suspicious about the condition of the back that I wrote to the mother after her return home and also to the medical attendant, and reiterated my fears of commencing bone disease. In the course of a few weeks, a distinct swelling appeared in the back, and the child was again brought up to Edinburgh to see me. On examination, I found a distinct but not large elastic swelling in the mid-dorsal region; there was still absolutely no tenderness on pressure and no spontaneous pain. I now

asked Professor Annandale to see the patient. He was definitely of opinion that the case was one of Pott's disease, although he agreed that the entire absence of pain was most exceptional, and that caries of the vertebræ very rarely indeed occurs at such an early age. As this diagnosis necessitated surgical treatment, the case was transferred to Professor Annandale's care. I did not again see the patient. Professor Annandale informs me that the swelling gradually increased in size and the patient became greatly emaciated. There was never any pain. It was obvious that the disease was sarcoma, not caries. A few days before death, a diagnostic incision was made into the tumour, more with the object of doing everything that it was possible to do than with the expectation of finding and evacuating pus. Nothing was evacuated. The incision showed that the swelling was sarcomatous. There was unfortunately no post-mortem.

I have described this case in considerable detail for it is of quite exceptional interest and rarity. It shows *firstly*, that compression myelitis may be rapidly developed as the result of sarcoma of the vertebræ without any pre-existing symptoms and signs of the bone disease; *secondly*, that for some time after the development of the myelitis there may be no evidence of bone disease; and *thirdly*, that the development of serious symptoms may result from a comparatively trivial injury—a slight twist or sprain of the back.

Diagnosis.—Compression myelitis is, as a rule, easily recognised. Transverse myelitis, especially when subacute or chronic, is the condition with which it is most likely to be confounded.

The differential diagnosis of compression myelitis and of ordinary transverse myelitis.—In most cases of compression myelitis the development of cord-symptoms is preceded or accompanied by symptoms and signs indicative of the primary disease, whatever it may happen to be, which is the cause of the compression, and by symptoms indicative of pressure on, or involvement of, the nerve roots. These preliminary symptoms are the most important facts which have to be taken into account in connection with the diagnosis. In the absence of such preliminary or associated symptoms, a diagnosis of compression myelitis is rarely if ever justifiable. In ordinary myelitis there is little or no pain in the back; and although a girdle sensation and ring of peripheral hyperæsthesia may be present, these symptoms are seldom so prominent as to suggest localised pressure upon nerve roots.

The differential diagnosis of the cause of the compression.—The

intra-dural causes of pressure myelitis, such as intra-dural tumours and pachymeningitis cervicalis hypertrophica, are very rare lesions. In the great majority of cases, then, the diagnosis as to the cause of the compression turns upon the presence or absence of an extra-dural cause; and since Pott's disease is by far the most common extra-dural cause, in every case in which the symptoms seem to indicate a compression myelitis, the greatest attention should be given to the condition of the spinal column and the presence or absence of any symptoms or indications of bone disease. As I have already said, the local signs of caries are usually distinct enough. In the rare and quite exceptional cases in which they are absent, the diagnosis is most difficult or impossible. In such cases, the age and constitutional condition of the patient, the hereditary history, the presence or absence of tubercular or malignant disease in other organs and the effects of treatment may give a clue to the true nature of the case, or at all events suggest the suspicion, as in the case of sarcoma to which I have directed your attention, that the case is not an ordinary simple case of myelitis.

Pott's disease may occur at any age, but it is usually met with in children (not infants) and in young adults. Malignant disease is much more frequent in the later periods of life, although sarcoma may, as we have seen, occur even in infancy.

Where there is no evidence of Pott's disease or of malignant disease, the possibility of the pressure being due to an aneurism (when the cause of the pressure appears to be extra-dural) must be remembered.

Prognosis.—In cases of compression myelitis, the prognosis chiefly depends upon the nature of the compressing lesion—whether it is curable or not—and upon the extent and severity of the myelitis. In cases of Pott's disease, the prognosis is, of course, very much more favourable than in cases of malignant disease of the bones. In the great majority of cases of Pott's disease in which there is no local abscess, the caries is curable provided that appropriate treatment is applied *for a sufficiently long period of time*. Malignant disease in this situation is certainly fatal. The importance of the nature of the primary disease is so obvious that I need say nothing more regarding it.

As regards the myelitis itself, we may say that, speaking

generally, a myelitis due to compression is much more likely to get well, provided the compression can be removed, than an ordinary transverse myelitis. In many cases of compression paraplegia, the paralysis is merely the result, or partly the result, of interruption of function and not of destruction of tissue. This is an important point in connection with the prognosis. But even granting that the paralysis is due to myelitis, there is, other things being equal, more hope of recovery than in ordinary transverse myelitis, always provided that the compression can be removed. The myelitis is, as a rule, less severe than in most cases of ordinary transverse myelitis; and a healthy cord in which myelitis is developed as the result of compression recovers (other things being equal) more satisfactorily than a cord in which myelitis has, so to speak, been established idiopathically. But this is a very general statement, for in some cases of myelitis due to compression the destruction is quite as severe as in many cases of ordinary transverse myelitis.

In cases of compression myelitis in which the paralysed muscles are markedly atrophied, in which the sphincters are paralysed and in which bed-sores develop the prognosis as regards recovery is, of course, much more unfavourable. But it is important to remember that even in those cases in which the bladder has been paralysed for months, the patient may ultimately get quite well. The following is a case in point:—

Case of spinal caries; suddenly developed paraplegia; paralysis of the bladder; ammoniacal urine and cystitis; formation of abscesses on each side of the spinal column in the lumbar region, due to the local pressure of a steel support; aspiration; complete cure after a year's rest in bed with a poroplastic support.

D. S., aged 21, was seen with Dr. Wood of Markinch on 27th May 1888.

The patient had suffered for many years from caries of the spine, for which he had worn a steel support. A few weeks before the paraplegia developed, he had been standing and walking about more than usual. This excessive strain was followed by pain in the back and then by rapidly developed loss of power in the legs.

When I saw him, the bladder and lower extremities were completely paralysed; the urine was ammoniacal and loaded with pus,

blood and vibrios. The bowels were obstinately constipated. The temperature was normal; the pulse numbered 92 in the minute. The patient was very restless. The knee-jerks were diminished. There was a marked spinal curvature with projection of the 7th, 8th and 9th dorsal vertebræ. The paralysis had developed two days before my visit.

The patient was put to bed and for some days continued much *in statu quo*.

On *June 2nd*, the pulse was 120; the temperature 101°. Profuse sweatings occurred at irregular intervals and there was tenderness and swelling on each side of the spine in the lumbar region. The patient also complained of cough and of a stinging pain in the lower part of the right chest.

On *June 10th*, the swelling in the back had increased and fluctuation could be detected in it. The pulse was 120; temperature 102°; respirations 28. The cough was very troublesome; the patient was spitting a good deal of muco-purulent material. Friction and fine crepitations were heard over the left base. Profuse sweating still continued. The paralysis of the legs was slightly less, the paralysis of the bladder much *in statu quo*.

On *June 16th*, Professor Chiene aspirated each of the abscesses; there was one on each side of the spinal column in the lumbar region. The two abscesses did not seem to communicate. The abscesses exactly corresponded to the points at which the base of the steel support, which the patient wore when he was going about, had pressed upon the back.

On *June 20th*, the back looked well; the pus had not reformed and the abscess cavities seemed to be contracting and consolidating. The temperature, pulse and respirations were normal. The paralysis of the legs was less; the patient also was getting more power over the bladder.

After this date there was slow but steady improvement.

On *June 12th*, it was noted:—The ankle-clonus is less marked; the bladder still remains paralysed; the general condition is satisfactory.

Towards the end of November, a poro-plastic jacket was fitted to the back. The patient had previously been lying on a water-bed. The patient was kept on the water-bed in the poro-plastic jacket until the 13th of May 1889; he was then for the first time allowed to get up and sit in a chair. A day or two after this, he took a few steps across the room. This exertion was followed by an attack of hæmoptysis which subsided satisfactorily under treatment.

At the end of June, the patient was allowed again to get up, and from this date he improved very rapidly and continuously.

On *October 23rd*, 1889, he walked into my consulting-room, saying that he was quite well. A few days previously he had walked three miles

and had not felt the least tired. There was no pain or swelling in the back. The knee-jerks were still somewhat exaggerated. The paralysis of the bladder, which had lasted for six months, had been completely recovered from. During the six months that the bladder remained paralysed, it had been washed out twice daily with a weak antiseptic; whenever the washing was discontinued, the paralysis of the bladder seemed to increase and the urine became ammoniacal and purulent.

The following are the notes of a second illustrative case of compression myelitis in which complete recovery also resulted.

Case of caries in the upper cervical vertebræ; paralysis of all four limbs; paralysis of the bladder; vasomotor œdema; complete recovery after a year's rest in bed.

Master Y., aged 7, was seen with Dr. Kidd of Alyth on 28th May 1890.

A year previously the patient had been threatened with tabes mesenterica, but had completely recovered. For some two months before my visit, he had complained of pain and stiffness in the neck and of severe neuralgic pains in the neck and head. A week before my visit, the upper limbs had become paralysed.

On examination, I found complete paralysis of the upper extremities, very marked but incomplete paralysis of the legs and complete paralysis of the bladder. Both the upper and lower extremities were swollen; the œdema was so marked that I suspected the presence of Bright's disease, but the urine contained no albumen; it was highly phosphatic. The œdema was obviously due to vasomotor paralysis. A swelling was present in the upper part of the neck at the junction of the spine with the head; the slightest movement caused intense pain; the swelling was exceedingly tender on pressure.

It was arranged to apply a rigid support to the spine and to keep the patient at absolute rest in bed. He was unable, however, to bear the pressure of the support; his head was therefore fixed by pillows. Tincture of iodine was painted over the swelling and iodide of potassium was given internally. Absolute rest was most scrupulously carried out; the boy was a splendid patient and did exactly as he was told. Gradual improvement took place.

At the end of seven weeks, some slight return of motor power was observed in the arms. The paralysis of the legs and bladder began to disappear at the end of five weeks.

In July 1890, he was lifted on to a specially-made couch with a head-piece which accurately fitted the head and neck and which prevented any movement. He lay on this couch during the day; his head was supported by pillows at night.

On *October 6th*, 1890, he was able to write me a letter to say that he was very much better.

The patient was kept at absolute rest, either on the couch or in bed, for a whole year. On *May 28th*, 1891, he was allowed to get up for the first time.

On *June 8th*, 1892, he came to see me in Edinburgh. He was then quite well, running about actively. All trace of paralysis had disappeared. The 3rd cervical vertebra was slightly prominent. There was no pain in the neck; the movements of the head towards the right and in a backward direction were slightly restricted, but absolutely unattended with pain.

The patient has since remained quite well.

Treatment.—The essential objects of treatment are to remove compression from the cord, to cure the primary lesion, and to endeavour to promote repair in the damaged cord. In those cases in which root-symptoms are prominent, the pain must of course be relieved by appropriate remedies.

In cases of Pott's disease of the vertebræ with compression myelitis or other nerve symptoms (root-symptoms), the treatment is partly local and partly constitutional.¹ Local rest is the first essential. The patient should be kept at absolute rest in bed for several months at least. I prefer myself to place such patients on a water-bed and in a rigid support. A poro-plastic jacket, which is accurately moulded to the body so as to form a comfortable and rigid support for the spinal column should be continuously worn. The parts of the jacket which cover the diseased bone (supposing that the caries is situated in the dorsal region, and this is the usual position), the armpits, hips, iliac spines, etc., should not be hardened, but should be allowed to remain soft and pliable. A water-bed is, in my experience, very advantageous; most patients can lie comfortably on the back on a water-bed in a rigid support for weeks or months, I might almost say for years. It is advisable to have two jackets, and to change the jacket once a week; that, I think, is sufficient. In changing the jacket great care must be taken that the diseased spine is not strained or twisted.

In those cases in which symptoms of pachymeningitis are

¹ Cases of Pott's disease in which there are no nerve symptoms belong to the surgeon; but in these cases, the treatment of the bone disease is of course the same.

prominent, counter-irritation, especially the application of the actual cautery, is often advisable. Blistering and iodine in some cases also do good. You must remember that if the patient is to lie flat on his back in a rigid support, it is difficult to apply counter-irritation satisfactorily. Hence, it is usually advisable to try what rest and support, without counter-irritation, will do, at all events in the first instance.

If there is reason to suppose that a distinct abscess, connected with the diseased bone, is exerting pressure on the spinal cord and producing the paraplegia, the advisability of surgical interference (aspiration, incision, laminectomy, etc.), must of course be considered. Though this is a surgical question, I may say that, from what I have seen of trephining the spine, I have come to regard the operation as a serious one. In cases of Pott's disease it is, so far as my experience enables me to judge, rarely advisable. It should seldom, if ever, I think, be performed until a prolonged trial has been given to the rest-poroplastic-water-bed plan of treatment. I am speaking of those cases in which there is no distinct local abscess. The great majority of cases of caries of the vertebræ can be cured by rest, provided that the rest is sufficiently prolonged. It is, in many cases, necessary to keep the patient at absolute rest for a year at least. Provided this is done, the paralysis will usually disappear, always granting of course that the tissues of the cord are not destroyed and disorganised by myelitis. It is unnecessary to say that in those cases in which the tissues of the cord are destroyed operative procedure is useless so far as the paralysis is concerned.

In addition to the local (rest) treatment, the general health must of course be maintained in the best possible state of efficiency. The greatest attention should be given to the ventilation of the sick-room. The patient must be well and judiciously fed. Tonics, cod-liver oil, maltine, quinine, syrup of the iodide of iron, the compound syrup of the phosphates, etc., should be administered.

If the bladder is paralysed, great care should be taken to prevent cystitis. If septic cystitis develops, the bladder should be washed out daily with a solution of boracic acid, and boracic acid (twenty grains three times daily in the case of an adult) should be given by the mouth.

In those cases in which spastic symptoms are prominent,

dilute hydrobromic acid, or a mixture containing dilute hydrobromic acid and quinine, is often beneficial.

As the bone lesion becomes quiescent and the pressure of the cord lessens, the nutrition of the limbs demands attention. Judiciously employed, massage, electricity, and, in some cases, strychnine or nux vomica, are useful at this stage. When the exaggeration of the deep reflexes continues, hydrobromic acid is a valuable remedy. In those cases in which the exaggeration of the knee-jerks and the presence of ankle-clonus are associated with a marked degree of muscular atrophy, a small dose of nux vomica or strychnine may be combined with the hydrobromic acid, the effect (of the nux vomica or strychnine) being very carefully watched.

LECTURE XXXIV

PACHYMEMINGITIS INTERNA HÆMORRHAGICA

IN addition to the form of pachymeningitis which we considered in the last lecture, two other varieties must be described. In both of them, the internal surface of the dura mater is affected. Both are very rare. The first is termed pachymeningitis interna hæmorrhagica.

Morbid Anatomy.—In this condition the lesion is identical with that of pachymeningitis interna hæmorrhagica cerebialis. The cranial variety is far more common than the spinal.

On naked-eye examination, the internal surface of the dura mater is found to be covered with a layer of chocolate or rust-coloured inflammatory exudation or blood clot in various stages of organisation. In many cases, there are several layers, which appear to have been formed at different dates. The inflammatory exudation and hæmorrhagic extravasation may be either limited to localised areas of the dura mater or scattered more or less extensively over the whole of its inner surface. Recent extravasations of blood are often present, and cyst-like cavities containing clots or collections of serum are occasionally found.

Different opinions are held as to the exact nature of the lesion. According to one view, the hæmorrhagic extravasations are poured out from delicate, thin-walled vessels formed in the midst of an inflammatory exudation. This view supposes that an inflammation of the dura mater is the first event—the primary lesion. According to another view—and I confess that this seems to me the more probable supposition—the hæmorrhage is the first event and the inflammatory lesions are secondary, i.e. developed round the blood clots.

On microscopical examination, numerous thin-walled vessels can usually be seen in the organised parts of the exudation.

Etiology.—Like hæmatoma of the dura mater, the condition is usually met with in persons who have been addicted to alcoholic excess or who are suffering from general paralysis or other forms of chronic insanity. A traumatic form has also been described.

Clinical History.—In the great majority of cases the condition is, as I have already stated, associated with hæmatoma of the cerebral dura mater. In cases of this kind, the cerebral symptoms usually occupy the chief place in the clinical picture, and are apt to entirely overshadow those which are due to the affection of the spinal dura mater.

The spinal symptoms are those of chronic and ill-defined meningitis, such as:—Pain in the back; stiffness of the spine; and slight root-symptoms indicative of motor and sensory irritation or of motor and sensory impairment in localised sensitive and muscular areas. In those cases in which a considerable amount of blood is suddenly poured out from the thin-walled vessels in the inflammatory exudation, symptoms suggestive of meningeal hæmorrhage may be developed; but it is very rarely that these symptoms are sufficiently characteristic to enable the condition to be recognised with any degree of probability during life.

Diagnosis.—The presence of pachymeningitis interna spinalis should be suspected when symptoms of chronic meningitis are associated with symptoms suggestive of cerebral hæmatoma. If, in addition, sudden exacerbations of the spinal symptoms every now and again occur (symptoms suggestive of repeated hæmorrhagic extravasations) a more positive diagnosis may be ventured upon.

Prognosis.—If the condition could be positively diagnosed, the prognosis would necessarily be very grave; for the cerebral variety of pachymeningitis interna hæmorrhagica (with which the spinal form is almost always associated) is almost invariably fatal—or perhaps it would be more correct to say that the cerebral lesions with which non-traumatic hæmatoma of the cerebral dura mater is associated are almost invariably fatal.

Treatment.—This is identical with the treatment of pachymeningitis interna hæmorrhagica cerebialis. I shall return to the subject at a subsequent period of the course.

PACHYMENINGITIS INTERNA HYPERTROPHICA

Morbid Anatomy.—This affection, which has been described by Professor Charcot under the term *pachyméningite cervicale hypertrophique*, seems to consist in a chronic inflammation of the internal surface of the dura mater. The essential feature of

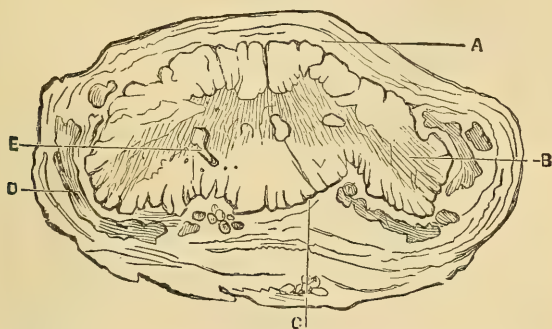


FIG. 166.—Transverse section of the cervical enlargement of the spinal cord in a case of hypertrophic cervical pachymeningitis.—(After Jaffroy.)

A, hypertrophied dura mater. B, nerve roots traversing the thickened meninges. C, pia mater confounded with the dura mater. E, section of two canals newly excavated in the grey substance.

the lesion is the enormous amount of cicatricial and fibrous thickening which is produced. The soft membranes are involved, and the thickened tissue extends in the form of a ring round the spinal cord (see fig. 166). The nerve roots arising from the cord at the seat of the lesion are first irritated and then destroyed, and the cord itself is subjected to slow compression. In consequence of this compression, inflammatory changes characteristic of pressure myelitis are developed, and the tissues of the cord become more or less disorganised.

In the great majority of cases the lesion is situated in the cervical region.

Clinical History.—Charcot described two stages in the course of the disease, viz. :—(1) a stage of irritation which usually lasts for two or three months ; and (2) a stage of subsequent paralysis and muscular atrophy.

First stage.—The symptoms characteristic of the first stage are due to irritation of the posterior and anterior nerve roots which pass through the affected portion of the membranes. They consist of:—(a) fixed pain in the neck or back and hyper-æsthesia in the sensitive areas of the affected nerve roots ; the

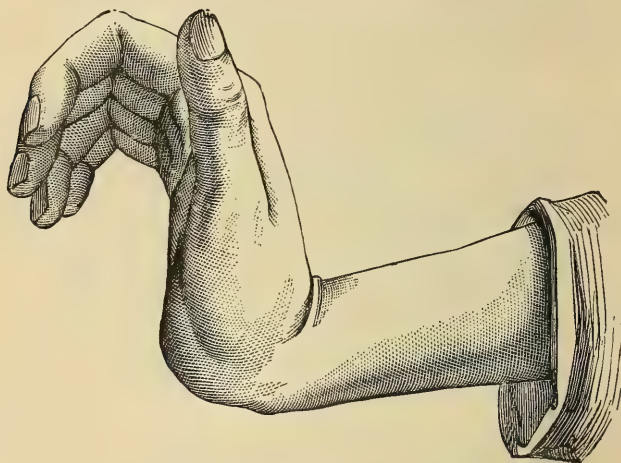


FIG. 167.—*The position of the hand in pachymeningitis cervicalis hypertrophica, affecting the lower part of the cervical enlargement.*—(After Charcot.)

pains are often very severe and are felt in the back, shoulders, arms, or upper part of the thorax ; and (b) muscular twitchings and spasms in the muscles supplied by the irritated anterior nerve roots (rigidity of the neck and upper parts of the spine, and of the muscles of the upper extremities).

In addition, some symptoms indicative of sensory and motor impairment (some motor paralysis and anæsthesia) are usually present.

Herpetic eruptions and trophic disturbances in the skin may also be developed during this, the first, stage of the disease.

Second stage.—After the symptoms of the first stage have lasted for two or three months, the second stage is gradually entered upon. It is characterised by symptoms of sensory and

motor impairment (anaesthesia, paralysis and muscular atrophy) distributed in the sensory and muscular areas of the nerve roots which are involved, and of symptoms due to pressure on the spinal cord.

The faradic excitability of the paralysed and atrophied muscles gradually becomes diminished or abolished, and characteristic contractures and deformities are in many cases developed.

When the lesion involves the nerve roots arising from the

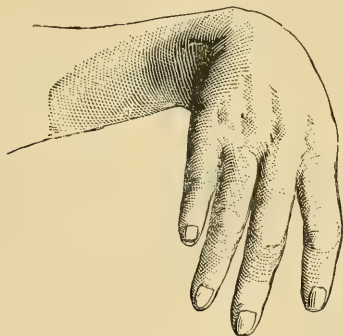


FIG. 168.—*The position of the hand in pachymeningitis cervicalis hypertrophica, affecting the upper part of the cervical enlargement.*—(After Ross.)

lower end of the cervical enlargement (the last cervical and first dorsal segments), the small muscles of the hand and the muscles of the forearms supplied by the ulnar and median nerves are chiefly affected, and the hands are apt to assume the position shown in fig. 167.

When the lesion is situated a little higher up (6th and 7th cervical segments), the extensor muscles of the forearms supplied by the musculo-spinal nerve are affected, and the hand is apt to assume the position shown in fig. 168.

When the lesion is situated still higher up (4th and 5th cervical segments), the muscles of the upper arm and shoulder girdle are affected.¹

In consequence of the compression of the spinal cord, myelitis at the seat of the compression is, as I have already said, usually

¹ In the table (page 608) the distribution of the nerves attached to the different segments of the cervical portion of the spinal cord is shown.

developed. Descending degeneration in the motor tracts below the lesion and a condition of spastic paraplegia result. When the compression is severe, the sensory conduction from the parts of the body below the lesion may also be interfered with; in short, the condition of the lower limbs may exactly simulate that which results from a transverse myelitis.

Course and Duration.—The course is usually slow. Some cases terminate in death, the fatal issue being usually due to cystitis, bed-sores, intercurrent complications, etc. In other cases, the morbid process seems after a time to be arrested. The paralysis and rigidity in the lower limbs may be partly recovered from, but the contractures and deformities in the upper extremities usually remain.

Diagnosis.—Owing to the localised atrophy in the muscles of the upper limbs, the condition may, at first sight, be mistaken for amyotrophic lateral sclerosis or progressive muscular atrophy. But the resemblance is only superficial.

The differential diagnosis of pachymeningitis cervicalis hypertrophica and amyotrophic lateral sclerosis.—In both conditions, there may be rigidity and atrophy in the upper extremities, with exaggeration of the deep reflexes in the upper and lower limbs. But there is no real difficulty in the diagnosis. The two affections are at once distinguished by the facts:—(1) That in pachymeningitis cervicalis hypertrophica, symptoms indicative of irritation of the motor and sensory roots at the seat of the lesion are conspicuous features. (2) That in amyotrophic lateral sclerosis, the muscles of the lower extremity, after being for a time rigid, undergo marked atrophy; while, in pachymeningitis cervicalis hypertrophica, although the muscles of the lower limbs may be paralysed and in a condition of spastic paraplegia, marked atrophy does not occur. (3) That in amyotrophic lateral sclerosis the course is usually pretty rapid and continuous and the morbid process often involves, or extends to, the medulla oblongata; while in pachymeningitis cervicalis hypertrophica the progress is almost invariably slow, arrest and incomplete recovery are comparatively common, and the lesion shows little or no tendency to extend upwards and to invade the medulla oblongata.

The differential diagnosis of pachymeningitis cervicalis hypertrophica and progressive muscular atrophy.—In the second stage, when the muscular atrophy in the upper extremities is marked, pachymeningitis cervicalis hypertrophica may in some degree resemble progressive muscular atrophy; but the two diseases are easily distinguished by the following facts:—(1) The pronounced sensory disturbances (sensory root-symptoms), which are so characteristic of pachymeningitis cervicalis hypertrophica, are completely absent in progressive muscular atrophy. (2) The fact that in pachymeningitis cervicalis hypertrophica, the muscular atrophy is preceded by a stage of muscular rigidity and tension. (3) The condition of the lower extremities. In the second stage of pachymeningitis cervicalis hypertrophica, spastic paraplegia is a marked feature; but in ordinary typical cases of progressive muscular atrophy, although the knee-jerks are often exaggerated, distinct spastic symptoms (rigidity and paralysis) are not developed in the lower extremities.

The differential diagnosis of pachymeningitis cervicalis hypertrophica and other conditions which produce compression myelitis.—From the statements which I have just made, you will easily understand that root-symptoms and symptoms indicative of compression myelitis are, from a diagnostic point of view, the most characteristic features of pachymeningitis cervicalis hypertrophica. These symptoms clearly show that the lesion is extra-medullary in position.

Having arrived at this conclusion, it is often a difficult matter to make the next step in the diagnosis, viz., to determine the exact pathological character of the extra-medullary lesion which is present. The differential diagnosis in most cases lies between Pott's disease of the vertebræ with associated pachymeningitis externa, extra-medullary tumours involving the spinal bones or membranes, and pachymeningitis cervicalis hypertrophica. In trying to determine which of these conditions is the cause of the symptoms, the most important points which have to be taken into account are:—The condition of the spinal column (the presence or absence of local pain, swelling and other indications of bone disease); the severity of the root-symptoms; the locality of the lesion; the age of the patient; the condition of the temperature (in some cases of Pott's disease a suppurative temperature is present); the rapidity with which the symptoms are developed; and

the presence or absence of lesions in other organs indicative of scrofula, cancer, or sarcoma.

In cases of pachymeningitis interna hypertrophica, the patient is usually middle-aged; except in some of the cases in which the condition is the result of traumatic injury, there is no prominence of the spinal bones and no external swelling; the local pain in the back (neck) and the root-symptoms are usually very prominent and are bilaterally distributed; the lesion is almost always situated in the cervical region, all four limbs are, therefore, in most cases involved; the paralysed muscles in the upper extremities are markedly atrophied; the temperature is normal; there are no indications of tubercle or malignant disease in the other organs or tissues; and the mode of development and course of the symptoms are slow and gradual.

Treatment.—The same measures of treatment which have been recommended in cases of pachymeningitis externa should be employed. The more important are :—Rest in the recumbent position on a water-bed; counter-irritation by the actual cautery at the seat of the lesion; the internal administration of (a) iodide of potassium and mercury (with the object of producing absorption of the inflammatory products), (b) hydrobromic acid or bromide of potassium to allay spasm, and (c) morphia to allay pain; massage and passive movements to prevent and remove contractures and deformities; after the spasm and rigidity are allayed and removed, the faradic current, massage, and small doses of strychnine or nux vomica and arsenic to promote and restore the nerves and muscular nutrition; and attention to the general health (good feeding, tonics such as quinine) and to the condition of the bladder and rectum.

INTRA- AND EXTRA-MEDULLARY SPINAL HÆMORRHAGE

Introductory remarks.—Hæmorrhage into the substance or on to the surface of the spinal cord, unless merely microscopic in amount, is usually due to traumatic injury; it is very rarely the result of disease. In this respect, there is a striking contrast between the diseases of the spinal cord and the diseases of the brain. When we come to the diseases of the brain, we shall see that coarse vascular lesions of all kinds, and particularly extravasations of blood into the substance of the brain, are by far the most common and important acute diseases of the brain which occur in the adult. It is very different in the case of the spinal cord; nevertheless, blood may be poured out either into the substance or on to the surface of the spinal cord. The former we term *intra-medullary*, and the latter *extra-medullary* hæmorrhage.

INTRA-MEDULLARY HÆMORRHAGE

Morbid Anatomy and Etiology.—The same conditions which tend to produce extravasations of blood into the substance of the brain may lead to the production of extravasations of blood into the substance of the spinal cord (spinal apoplexy). The two great causes are:—(1) alterations in the vascular walls which weaken their resistance; and (2) increased blood pressure.

As I have just remarked, spinal apoplexy is a rare condition, and when it does occur it is usually the result of some pre-existing disease, such as a gliomatous condition of the cord tissues. The low blood pressure in the long and relatively speaking small arteries which supply the spinal cord with blood prevents the production of atheroma, periarteritis and miliary aneurisms, and lessens the risk of rupture even if the arteries should happen to be diseased. Small extravasations are, as we have seen, by no means uncommon in acute myelitis.

Cases are occasionally met with in which cord symptoms are suddenly or very rapidly developed after the sudden arrest of menstruation. It is reasonable, I think, to conclude that in some

cases of this kind the lesion is a hæmorrhage. Hæmorrhage into the substance of the cord may also, of course, result from traumatic injury, such as a direct blow on the spine, a fall on the feet, etc. It is probable that in many of the cases in which organic disease of the spinal cord follows an injury to the back and in which the spinal bones are uninjured the organic lesion has its starting-point in minute capillary hæmorrhages into the grey matter. Large extravasations of blood sufficiently extensive to produce immediate symptoms and to destroy life are seldom the result of traumatic injury except in those cases in which the spinal bones are fractured or dislocated. In cases in which the spinal bones are injured, extensive extravasations on the surface of the cord and bruising with ecchymosis of the cord itself are, of course, very common; but large hæmorrhages into the substance of the cord, without associated blood extravasations on the surface rarely result from traumatic violence.

Spinal apoplexy may occur at any period of life, but it seems to be more frequent during youth and early adult life than at other periods. In this respect, it differs very markedly from cerebral hæmorrhage, which is far more frequent in middle-aged and old people. The earlier occurrence of spinal apoplexy is probably explained by the fact that in many cases the bleeding is not a primary, but a secondary and, as it were, an accidental event occurring in the course of some other morbid process, such as a glioma.

The amount of blood which is extravasated is usually small; the clot is seldom larger in size than an almond; the blood is almost always poured out into the central grey matter. The delicate nerve tissue is ploughed up, and the function of the spinal cord suddenly interrupted. A sudden extravasation of blood, even of small size, is usually followed by the immediate production of paralysis of all the muscles supplied by nerves passing off below the lesion. Should the patient survive for a sufficiently long period of time, inflammatory changes may occur in the neighbourhood of the clot and secondary degenerations may be developed.

In some cases, the extravasation takes place so rapidly that the patient is struck down with immediate paralysis of a paralytic type; sudden pain in the back is in many cases experienced; but there is usually no loss of consciousness. In other cases in

which the blood is poured out more gradually, the full development of the paralysis may not be reached for some hours; in cases of this kind, pain in the back may be slight or altogether absent.

The motor, sensory, reflex, vesical, rectal and other derangements may be identical with those which result from an ordinary acute transverse myelitis; practically speaking, a large hæmorrhagic extravasation into the substance of the cord may be regarded as an acute transverse lesion.

Diagnosis.—The only conditions with which spinal apoplexy is likely to be confounded are poliomyelitis anterior acuta, acute myelitis, and extra-medullary hæmorrhage, in all of which rapidly developed paraplegia may be produced.

The differential diagnosis of intra-medullary hæmorrhage and of poliomyelitis anterior acuta presents no difficulty. In cases of primary hæmorrhage, the onset of the paralysis is, as a rule, much more rapid than in poliomyelitis anterior acuta, and is not attended with, or preceded by, fever. In cases of spinal apoplexy, severe pain in the back is often complained of, sensory disturbances (anæsthesia below the seat of the lesion) are usually prominent, the bladder and rectum are generally paralysed, and in those cases in which the patient survives bed-sores are often developed. If the patient should survive for a sufficient length of time, the paraplegia which results from an intra-medullary hæmorrhage resembles that characteristic of an ordinary transverse myelitis rather than of poliomyelitis anterior acuta; it is only those muscles connected with the grey matter which is destroyed which undergo rapid atrophy and present the reaction of degeneration. Further, an intra-medullary hæmorrhage of large extent is almost always fatal and often rapidly fatal; whereas it is only in the most exceptional instances that poliomyelitis anterior acuta results in death.

The differential diagnosis of intra- and extra-medullary hæmorrhage may be a matter of great difficulty. The point of chief diagnostic importance is that in intra-medullary hæmorrhage (as in acute myelitis) symptoms of motor and sensory impairment (paralysis) are rapidly or suddenly developed, irritative symptoms (pain in the back excepted) being absent or merely slight and temporary; whereas in extra-medullary hæmorrhage (as in acute meningitis)

symptoms of motor and sensory irritation are, relatively speaking, more marked. In cases of intra-medullary hæmorrhage, pain in the back is seldom so marked and so widely diffused as in extra-medullary hæmorrhage. Paralysis of the bladder, bed-sores and trophic alterations in the skin are more frequently produced by intra-medullary than by extra-medullary hæmorrhage. Much, however, depends upon the position and extent of the extravasation and the duration of the case. A small hæmorrhage on to the surface of the cord may produce comparatively few symptoms; while a large hæmorrhage which is rapidly poured out is usually followed by the rapid production of paraplegia; but even in cases of large extra-medullary hæmorrhage, the paralysis is usually preceded by well-marked symptoms indicative of sensory or motor irritation.

Prognosis.—The prognosis depends upon the size and position of the clot. Other things being equal, a large hæmorrhage is, of course, much more serious than a small one. A hæmorrhage of large size usually proves rapidly fatal. A small hæmorrhage in the upper part of the cervical enlargement usually kills rapidly in consequence of the paralysis of the respiratory muscles which it produces. A hæmorrhage into the lower part of the lumbar enlargement, which almost of necessity produces paralysis of the sphincters, is, other things being equal, more serious than a hæmorrhage into the dorsal region. The development of bed-sores is of grave significance. From these statements, you will readily understand that in those cases in which the patient survives the first effects of the hæmorrhage, the prognosis must, as in cases of acute myelitis, be guided by the exact character, extent and severity of the symptoms.

Treatment.—An attempt should be made in the earlier stages to arrest the bleeding. Provided that the nature of the condition is suspected or diagnosed, the same measures of treatment which have been previously recommended for the treatment of acute myelitis and poliomyelitis anterior acuta in their early stages should be adopted. The patient should be placed in the prone position; rest should be enjoined; ice-bags, leeches, or cups applied over the seat of the supposed hæmorrhage; and full doses of ergotine given subcutaneously. In theory this

treatment is admirable, but in practice it is rarely possible to carry it out sufficiently early to produce any decided effect upon the hæmorrhage itself. In most cases, even supposing that the symptoms are sufficiently definite to warrant a correct diagnosis—and I may say that I have never yet seen a case in which I had reason to suppose that a primary intra-medullary hæmorrhage had just occurred—the damage, as in poliomyelitis anterior acuta, is already done before the case comes under observation. Under such circumstances, all that the physician can do is to attempt to mitigate the after-effects of the lesion by reducing and keeping in check the inflammatory changes which develop around the clot. When the hæmorrhage is considerable, treatment, however active, is not likely to be attended with much success; cases of this kind rarely survive more than a few days. Should the patient live longer, the subsequent treatment of the case is the same as that of acute myelitis.

EXTRA-MEDULLARY HÆMORRHAGE

Extravasation of blood into the spinal membranes (spinal meningeal hæmorrhage) is, like intra-medullary hæmorrhage, a very rare condition except as the result of traumatic injury. I have already referred to those cases in which the hæmorrhage is due to pachymeningitis hæmorrhagica. In extremely rare instances, spinal meningeal hæmorrhage is due to the rupture into the spinal canal of an aneurism of the thoracic or abdominal aorta. It may also occur in the course of purpura, scurvy, and other conditions in which there is a tendency to blood extravasation. In some cases, blood which has been poured out into the cavity of the cranium, as the result for example of the rupture of an aneurism of one of the large vessels at the base of the brain, finds its way into the upper part of the spinal canal; in such cases, the spinal hæmorrhage is of no importance and is rarely if ever attended with any definite symptoms.

In some cases of spinal meningeal hæmorrhage, the blood is poured out between the dura and the bones, that is to say, on the outer surface of the dura; in others, between the dura and the soft membranes.

The amount of blood varies considerably in different cases.

It is sometimes sufficiently copious to envelop the whole cord. The blood may be partly fluid; when considerable it is generally dark-coloured and partly clotted.

Onset, Symptoms and Course.—In those cases in which the hæmorrhage is large and in which the extravasation occurs rapidly, initial symptoms indicative of meningeal irritation (*viz.*, pain in the back, shooting pains in the limbs or trunk, muscular cramps, twitchings, spasms, etc.) are speedily followed by symptoms indicative of motor and sensory impairment (paraplegia). In those cases in which the blood is slowly extravasated, the paralytic symptoms are more gradually developed. Fever and other symptoms indicative of meningitis are usually developed during the subsequent stage of reaction.

Diagnosis.—The more important symptoms of extra-medullary hæmorrhage from a diagnostic point of view are the sudden occurrence of symptoms indicative of meningeal irritation and the absence of fever at the commencement of the attack. When the hæmorrhage is considerable in amount and rapidly poured out, the distinction from intra-medullary hæmorrhage may, as I have already told you, be impossible. The chief points of difference have already been considered. (See page 593.)

The differential diagnosis of meningeal hæmorrhage and meningitis.—Meningeal hæmorrhage is distinguished from acute meningitis by the rapidity with which the symptoms are developed, by the absence of fever at the commencement of the attack, and by the more rapid development of paralysis.

Prognosis.—This is more favourable than in cases of intra-medullary hæmorrhage. In some cases, the symptoms gradually abate, and, if the hæmorrhage is small in amount, the patient may perhaps eventually get quite well. When the hæmorrhage is extensive, and especially when the upper part of the spinal cord is pressed upon, a fatal issue is apt to result. When the hæmorrhage is copious, the cord is apt to be compressed; myelitis may under such circumstances develop, and death may result from exhaustion, bed-sores, cystitis, etc.

Treatment.—The same method of treatment which has been already recommended for the arrest of an intra-medullary hæmorrhage should be adopted.

During the subsequent stages, the case should be treated as one of acute meningitis. Morphia may be required to relieve pain; and chloral and bromide of potassium to relieve spasm.

INTRA- AND EXTRA-MEDULLARY SPINAL TUMOURS

LET me next direct your attention to the new formations which affect the spinal cord, either directly or indirectly. Tumours arising within the spinal cord (intra-medullary tumours) are rare. Tumours which originate on the surface of the cord or in the spinal bones, though also rare, are somewhat more common. Extra-medullary tumours are apt to compress the spinal cord and to produce myelitis.

INTRA-MEDULLARY TUMOURS

Morbid Anatomy.—Gliomatous, glio-sarcomatous and tubercular tumours are the most common forms of intra-medullary tumours. Syphilitic gummata are usually developed on the surface of the cord, but they occasionally originate in its substance. Secondary deposits of sarcoma or melanotic sarcoma are occasionally, though very rarely, found in the substance of the spinal cord. In short, in the great majority of cases, tumours which originate in the substance of the spinal cord are primary.

Intra-medullary tumours are, as a rule, solitary. They are usually of small size; but gliomatous tumours may (as we have seen in our study of syringomyelia) extend from the top to the bottom of the spinal cord. Gliomatous tumours are more frequently met with in the cervical region, while tubercular tumours seem in most cases to develop in the lumbar enlargement. (See fig. 169.)

New growths which originate in the spinal cord seem in the great majority of cases to have their starting-point in the embryonic tissue around the central canal or in the pia mater or the vascular offshoots which pass from the pia mater into the cord tissue. Tubercular tumours seem in many instances to originate in the grey matter.

In its early stages, an intra-medullary tumour is often localised and limited to one half of a spinal segment, but as it grows and increases it compresses and involves the other half, until finally the whole structure of the cord at the level of the tumour may be disorganised and destroyed. It is needless to

say that secondary degenerations, ascending and descending, are under such circumstances developed.

Etiology.—In rare cases, a blow on the spine or injury of the back seems to be the starting-point of the new growth; but in the great majority of cases, there is no obvious exciting cause.

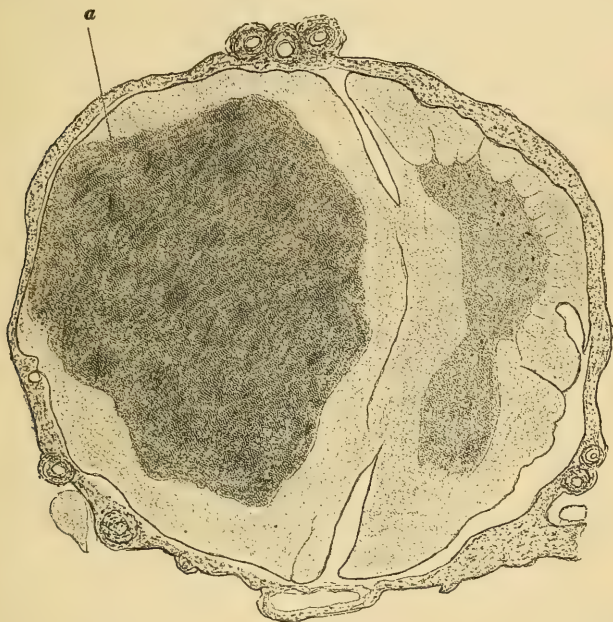


FIG. 169.—*Transverse section through the lumbar enlargement of the spinal cord, showing a large tubercular tumour occupying the greater part of the left half. (\times about 10 diameters.)*

The right half of the cord was very much compressed and partly degenerated; the right anterior horn of grey matter with its nerve cells is still distinguishable.

The tumour *a* is shaded dark.

Gliomatous tumours appear in many cases to be congenital. (See page 393.)

Onset, Symptoms and Course.—In some cases, the symptoms are, for a time, very indefinite and indistinct; in others, paralysis is developed at a comparatively early stage of the case. These differences depend upon the rapidity with which the tumour is growing and developing, and the effect which it produces upon the nervous structures. A slow-growing tumour may for a time

simply displace the nerve elements and cause atrophy of the parts in its immediate neighbourhood. In cases of this kind, the symptoms are ill-defined, and the tumour may, for a time, be latent. A rapidly-growing tumour may quickly destroy the delicate cord tissue or may quickly lead to the production of myelitis.

Pain in the back is in many cases experienced, but it is rarely a marked symptom; in this respect there is usually a striking contrast between tumours which originate in the spinal cord and tumours which are situated on the surface of the cord: in many cases of extra-medullary tumour the back pain is very severe.

The most marked symptom is usually motor paralysis. In the great majority of cases, the paralysis slowly develops and steadily increases. The extent and distribution of the paralysis vary, of course, with the position of the tumour and the extent of the transverse section of the cord which happens to be implicated. In the early stages, the paralysis may be unilateral; as the tumour extends, the paralysis not only becomes more marked, but usually affects the muscles on both sides of the body (i.e. produces paraplegia).

As I have already pointed out in describing the symptoms of syringomyelia, gliomatous tumours are in many cases attended with a peculiar derangement of sensation—loss of sensibility to heat and cold and to pain, the tactile sensibility being unimpaired.

Should the tumour involve the anterior cornu in the lumbar or cervical enlargements, localised muscular atrophy will of course be superadded to the paralysis. In the case of tubercular tumours, inflammatory changes (myelitis) are sooner or later developed round the new growth. The lesion then practically corresponds to a focal or transverse myelitis, the symptoms of which have already been described in full.

Tubercular tumours are usually, as I have already stated, situated in the lumbar enlargement of the cord. In cases of this kind, the paralysis, which may in the early stages be limited to one leg, after a time becomes paraplegic in distribution. Loss of the knee-jerks, marked atrophy in some of the paralysed muscles of the lower extremities, and paralysis of the sphincters of the bladder and rectum are often conspicuous symptoms in the later stages of cases of this kind.

In the case of gliomatous tumours, symptoms of spinal apoplexy are sometimes developed, the blood being extravasated from the thin-walled vessels of which the tumour in some cases largely consists.

From these statements, you will readily understand that the exact character of the symptoms depends upon the position of the tumour, its size, the rapidity with which it is growing, and especially upon any secondary changes in the spinal cord (compression, atrophy, myelitis, etc.) which it produces.

In the rare cases in which there is more than one tumour in the spinal cord, the symptoms may, of course, be much more complicated.

Diagnosis.—The diagnosis is often attended with great uncertainty, especially in the earlier stages. Leaving out of account the gliomatous cases in which as we have seen a positive diagnosis can often be made with considerable probability and in some cases with definite certainty, a premonitory stage of ill-defined symptoms, followed by symptoms indicative of a progressive focal lesion or chronic transverse myelitis should suggest the possibility of an intra-medullary tumour.

A paralysis beginning slowly and gradually in one leg or in one arm (i.e. in one lateral half of the spinal cord) and gradually extending to and involving the corresponding part (arm or leg) on the opposite side of the body, is highly suggestive of the presence of a tumour. The absence of pain in the back, of root-symptoms of an irritative kind (shooting pains and spasms), and of other symptoms indicative of an extra-medullary lesion make the diagnosis more certain. It must, however, be remembered that pain in the back is not an unfrequent result of intra-medullary tumours.

Prognosis.—In the great majority of cases, the prognosis is most unfavourable. To some extent, the opinion must be guided by the pathological character of the new growth. In syphilitic cases, recovery, which is usually incomplete, for some paralysis usually remains, may, of course, occur; but syphilitic tumours are very rarely intra-medullary in position. Tubercular tumours are possibly in rare instances recovered from; I am certainly not prepared to deny the possibility of recovery in these cases.

But practically speaking, in the vast majority of cases of intra-medullary tumour (syphilitic cases excepted), the prognosis is hopeless. In gliomatous cases, several years may elapse before death takes place.

Treatment.—Iodide of potassium or iodide of potassium and mercury should be thoroughly tried. When these remedies fail, arsenic may be prescribed. In tubercular cases, cod-liver oil, lacto-phosphate of lime and general tonics should be administered. Surgical interference is not advisable except perhaps in rare cases of syringomyelia. In the present state of our knowledge, the surgeon can hardly hope to successfully remove tumours which originate within the substance of the spinal cord itself. When the physician is in doubt as to the intra- or extra-medullary nature of the tumour, surgical interference may be advisable.

EXTRA-MEDULLARY TUMOURS

Morbid Anatomy.—Tumours springing from the bones, the fibro-cellular and fatty tissue between the dura and the bones, the membranes or nerve roots are more common than new formations of intra-medullary origin. The following are the chief varieties which are met with:—Inflammatory formations, cancerous, sarcomatous and syphilitic growths originating in the bones; sarcomatous, scrofulous, fibrous, myxomatous, syphilitic, fatty, cartilaginous growths and hydatid cysts originating in the membranes or between the dura mater and the bones; and sarcomatous, myxomatous, gliomatous, fibrous and neuromatous tumours springing from the nerve roots.

Extra-medullary tumours are usually localised and in most instances solitary. Sarcomata and neuromata are often multiple.

In fig. 170 a characteristic example of an extra-medullary tumour is represented.

Etiology.—As in the case of intra-medullary tumours, a history of traumatic injury to the spine, a blow on the back, etc.,

is in some cases forthcoming. Cancerous, sarcomatous and scrofulous tumours are sometimes secondary to similar diseased conditions in other parts or organs of the body; but this is exceptional.



FIG. 170.—*Compression of the cervical portion of the spinal cord by a tumour springing from an anterior nerve root. One and a half times the natural size.*

The cord is suspended by a string attached to it above the tumour.

The letter *A* points to the tumour which is attached to an anterior nerve root. The letter *B* points to a second nodule of new growth.

I am indebted to Dr. Banham of Sheffield and the late Dr. Goyder of Newcastle for the preparation from which the drawing was made.

Onset, Symptoms and Course.—The symptoms produced by extra-medullary tumours vary considerably in different cases; but speaking generally (a) pain in the back, (b) symptoms indicative of irritation or destruction of localised nerve roots and

(c) of compression of the spinal cord are the essential clinical characteristics.

In contrasting the extra- and intra-medullary tumours, the same general principle which has already been insisted upon in treating of other extra- and intra-medullary lesions (meningitis and myelitis) must be kept in view, viz., that in extra-medullary tumours, pain in the back and symptoms of irritative root-symptoms are usually prominent; whereas, in intra-medullary tumours, paralytic symptoms due to disorganisation of the tissues of the cord and myelitis are usually conspicuous.

In some cases, the clinical course may be divided into a first or irritative and a second or paralytic stage.

The first or irritative stage.—The rapidity with which the symptoms are developed is very variable. In some cases, the first stage, characterised by localised pain in the back, shooting pains, hyperæsthesia, slight sensory and motor impairment in the areas of the nerve roots which happen to be implicated, lasts for a considerable time.

The severity of the pain in the back is very variable. In some cases, as in the case of sarcoma in the child to which I have previously alluded, there is absolutely no back-pain; but this is exceedingly rare. In other cases, the pain is agonising. In the case of a man who was sent to me two or three years ago by Dr. Ballantine of Dalkeith, intense pain in the upper part of the neck was complained of. All four limbs gradually became paralysed; shooting pains in the upper limbs and marked atrophy with some rigidity of the muscles of the upper extremities were developed. In the lower limbs (which were partially paralysed) the knee-jerks were greatly exaggerated, and there was ankle-clonus. Although I had the opportunity of seeing the patient on only one occasion, the symptoms seemed to be sufficiently characteristic to warrant a diagnosis of an extra-medullary tumour. The grounds for this opinion were fully explained to the students at my Clinique and the question of operative procedure was considered. As I have no male beds in the Infirmary, the patient had to be admitted under the care of another physician. I had no opportunity of seeing the case again, but I understand that he died in the course of a short time, and that a tumour, the presence of which I had suspected, was found post mortem.

The exact distribution of the root-symptoms depends of course upon the position of the tumour and the number of nerve roots which happen to be implicated. In some cases, the nerve roots on one side only are involved; the root-symptoms are then of course unilateral in distribution. In other cases, the symptoms of the first stage are indicative of pressure on the cord rather than of irritation of the membranes or nerve roots.

The second or paralytic stage.—After the primary stage has lasted for a longer or a shorter period, paralytic symptoms are developed. Like the irritative symptoms, they may be due either to pressure on and destruction of the nerve roots, or to pressure on the spinal cord itself and the production of compression myelitis.

When the tumour is situated in the cervical region, the root-symptoms are of course distributed in the upper extremities.

All four limbs may be paralysed. The muscles of the upper extremities which, in the earlier stages, may be rigid, may ultimately become markedly atrophied. The paralysis in the lower extremities corresponds to that which is due to compression myelitis. When the compression of the cord is severe or the pressure myelitis considerable, the ordinary symptoms of transverse myelitis (motor and sensory paralysis below the lesion, exaggeration of the deep reflexes in the lower extremities, etc.) are developed. Tumours involving the cervical region may produce alterations in the size of the pupils.

When the tumour involves the mid-dorsal or lower dorsal region of the cord, the upper limbs are of course unaffected. The lower limbs are alone paralysed; their deep reflexes are exaggerated. The paralysed muscles in the lower extremities do not undergo marked atrophy.

When the tumour involves the lumbar enlargement, the lower limbs are paralysed. Some of the muscles of the lower extremities are usually markedly atrophied. The condition of the deep reflexes is variable; in some cases they are exaggerated; in others abolished. The condition of the reflexes of course depends upon the position of the tumour and the effects which it produces upon the nerve roots and anterior horns of grey matter corresponding to the respective reflex centres. Tumours which press upon the lumbar enlargement are usually associated with marked

disturbance of the bladder and rectum. In these cases, too, bed-sores may be developed just as in ordinary cases of transverse myelitis.

An extra-medullary tumour which is situated in the lower part of the spinal canal and which involves the nerve strands of the cauda equina usually produces bilateral paralysis in the lower extremities and marked disturbance of the bladder and rectum. In cases of this kind, root-symptoms (severe neuralgic pains, hyperæsthesia, anæsthesia dolorosa, cramps and spasms, and after a time paralysis and localised muscular atrophy, abolition of the reflexes, and anæsthesia) are usually very prominent symptoms. In many cases there are bed-sores. The pressure of a tumour upon the cauda equina is one of the most important causes of painful paraplegia.

The distribution of the sensory paralysis which is usually present is variable. It depends of course upon the number and distribution of the nerve roots which are most severely implicated.

Whatever the position of the tumour, pain in the back is usually a prominent symptom.

After the second stage is reached, the case usually progresses more rapidly. The patient ultimately dies either from exhaustion, from bed-sores, from cystitis, or from paralysis of the muscles of respiration or intercurrent lung complications. Paralysis of the respiratory muscles is only of course produced when the tumour involves the upper dorsal or cervical regions. Bedsores and cystitis are chiefly developed in those cases in which the lumbar enlargement or the cauda equina are affected.

Diagnosis.—The diagnosis is in some cases comparatively easy, but often attended with much uncertainty, more particularly in the earlier stages.

Pain in the back and root-symptoms, gradually developed, together with symptoms indicative of slow compression of the spinal cord, suggest that the lesion is extra-medullary in position.

When these symptoms develop slowly and are unattended with fever, when the lesion appears to be of limited vertical extent, and when there is no evidence of Pott's disease of the spine and no history of a dislocation or fracture of the vertebræ,

the diagnosis of an extra-medullary tumour is warranted, provided that the hypertrophic form of pachymeningitis can be excluded.

When, in addition, there is any evidence of cancerous, scrofulous, hydatid or syphilitic disease in some of the other organs or parts of the body, the diagnosis of an extra-medullary tumour is much more certain, and the grounds for forming an opinion as to its pathological nature more clear.

Anatomical diagnosis.—The position of the tumour and its vertical extent are ascertained by observing:—(1) The exact position of the pain in the back. This in many cases is strictly localised, and affords corroborative evidence as to the exact position of the tumour. (2) The exact distribution of the root-symptoms, more especially of the neuralgic (shooting) pains, and of the localised muscular atrophy. The particular nerve roots which are implicated can in this way be ascertained. (3) The condition of the reflexes. (4) The distribution of the paralysis. (5) The height of the anæsthesia and the hyperæsthesia. (See table, page 608.) In the case of a unilateral lesion (i.e. a tumour pressing upon one side of the cord), the position and vertical extent of the anæsthetic band on the same side as the lesion should be carefully observed. (6) The presence of any local alterations in the spinal column, such as thickening or protrusion of the bones, is also of course important. It is important to remember that the spinal cord terminates at the lower border of the first lumbar vertebra, and that the greater number of the spinal nerves leave the spinal canal at a point considerably below their points of attachment to the spinal cord. Hence, in the case of intra- and extra-medullary tumours the actual position of the tumour is usually above the upper limit of the anæsthesia; it may be as much as two or three vertebræ above this point.

Pathological diagnosis.—The exact pathological character of the tumour is determined by observing:—(1) The presence of associated diseased conditions which are likely to give a clue to the nature of the lesion—the presence, for example, of malignant disease of the liver or other organs of the body would be a sufficient reason for supposing that the intra-spinal tumour was also malignant; the presence of other syphilitic lesions that the tumour was in all probability syphilitic. (2) The constitutional peculiarities and tendencies of the individual. (3) The family

LOCALISATION OF THE FUNCTIONS OF THE SEGMENTS OF THE SPINAL CORD.—(*After Starr.*)

| SEGMENT. | MUSCLES. | REFLEX. | SENSATION. |
|-----------------|---|--|---|
| II. and III. C. | Sterno-mastoid. Trapezius. Scaleni and neck. Diaphragm. | Hypochondrium (?). Sudden inspiration produced by sudden pressure beneath the lower border of ribs. | Back of head to vertex. Neck. |
| IV. C. | Diaphragm. Deltoid. Biceps. Coraco-brachialis. Supinator longus. Rhomboid. Supra and infra spinatus. | Pupil. 4th to 7th cervical. Dilatation of the pupil produced by irritation of neck. | Neck. Upper shoulder. Outer arm. |
| V. C. | Deltoid. Biceps. Coraco-brachialis. Brachialis anticus. Supinator longus. Supinator brevis. Rhomboid. Teres minor. Pectoralis (clavicular part) Serratus magnus. | Scapular. 5th cervical to 1st dorsal. Irritation of skin over the scapula produces contraction of the scapular muscles. Supinator longus. Tapping its tendon in wrist produces flexion of forearm. | Back of shoulder and arm. Outer side of arm and forearm, front and back. |
| VI. C. | Biceps. Brachialis anticus. Pectoralis (clavicular part) Serratus magnus. Triceps Extensors of wrist and fingers. Pronators. | Triceps. 5th to 6th cervical. Tapping elbow tendon produces extension of forearm. Posterior wrist. 6th to 8th cervical. Tapping tendons causes extension of hand. | Outer side of forearm, front and back. Outer half of hand. |
| VII. C. | Triceps (long head). Extensors of wrist and fingers. Pronators of wrist. Flexors of wrist. Subscapular. Pectoralis (costal part). Latissimus dorsi. Teres major. | Anterior wrist. 7th to 8th cervical. Tapping anterior tendons causes flexion of wrist. Palmar. 7th cervical to 1st dorsal. Stroking palm causes closure of fingers. | Inner side and back of arm and forearm. Radial half of the hand. |
| VIII. C. | Flexors of wrist and fingers. Intrinsic muscles of hand. | | Forearm and hand, inner half. |

LOCALISATION OF THE FUNCTIONS OF THE SEGMENTS OF THE
SPINAL CORD.—(After Starr.)

| SEGMENT. | MUSCLES. | REFLEX. | SENSATION. |
|----------------|---|---|--|
| I. D. | Extensors of thumb. Intrinsic hand muscles. Thenar and hypothernar eminences. | | Forearm, inner half. Ulnar distribution to hand. |
| II. to XII. D. | Muscles of back and abdomen. Erectores spinæ. | Epigastric. 4th to 7th dorsal. Tickling mammary region causes retraction of the epigastrium. Abdominal. 7th to 11th dorsal. Stroking side of abdomen causes retraction of belly. | Skin of chest and abdomen, in bands running around and downward corresponding to spinal nerves. Upper gluteal region. |
| I. L. | Ilio-psoas. Sartorius. Muscles of abdomen. | Cremasteric. 1st to 3rd lumbar. Stroking inner thigh causes retraction of scrotum. | Skin over groin and front of scrotum. |
| II. L. | Ilio-psoas. Sartorius. Flexors of knee (Remak). Quadriceps femoris. | Patella tendon. Striking tendon causes extension of leg. | Outer side of thigh. |
| III. L. | Quadriceps femoris. Inner rotators of thigh. Abductors of thigh. | | Front and inner side of thigh. |
| IV. L. | Abductors of thigh. Adductors of thigh. Flexors of knee (Ferrier). Tibialis anticus. | Gluteal. 4th to 5th lumbar. Stroking buttock causes dimpling in fold of buttock. | Inner side of thigh and leg to ankle. Inner side of foot. |
| V. L. | Outward rotators of thigh. Flexors of knee (Ferrier). Flexors of ankle. Extensors of toes. | | Back of thigh, back of leg, and outer part of foot. |
| I. to II. S. | Flexors of ankle. Long flexor of toes. Peronei. Intrinsic muscles of foot. | Plantar. Tickling sole of foot causes flexion of toes and retraction of leg. | Back of thigh. Leg and foot, outer side. |
| III. to V. S. | Perineal muscles. | Foot reflex. Achilles tendon. Overextension of foot causes rapid flexion; ankle-clonus. Bladder and rectal centres. | Skin over sacrum. Anus. Perineum. Genitals. |

history. A strong hereditary history of tubercle or cancer, in the absence of any direct evidence, would suggest these forms of new growth. (4) The effect of treatment. In a suspected syphilitic case, rapid improvement under iodide of potassium and mercury would confirm the syphilitic nature of the new growth.

Treatment.—The same measures which have been recommended for the treatment of intra-medullary tumours are to be adopted and the possibility of removing the new growth considered. It is only in rare cases that operative procedure is likely to be successful; for, in the first place, tumours on the surface of the cord which can be removed by the surgeon are exceedingly rare lesions; and, in the second place, the diagnosis is often attended with much difficulty. But the difficulties in diagnosis are disappearing; and, thanks to antiseptic surgery, operations can now be undertaken which were formerly considered unjustifiable.

I would advise an operation in any case (1) in which the symptoms were clearly indicative of an extra-medullary tumour; (2) in which there was no evidence to show that the tumour was malignant in nature; and (3) in which an antisiphilitic plan of treatment, vigorously carried out for three or four weeks, had failed to produce a distinct beneficial effect. I need hardly add (4) that an operation should only be attempted in those cases in which the new growth can be localised with reasonable probability.

It must, however, be remembered that, except in syphilitic cases, we cannot hope, with our present therapeutic measures, to cure or remove extra-medullary tumours. We are justified, therefore, in risking something in the way of operative procedure. On the other hand, it must not be forgotten that trephining the spine is a much more serious procedure than trephining the skull. The operations which I have seen have left such an unfavourable impression on my mind that I should hesitate to advise an operation for the removal of a supposed tumour on the surface of the spinal cord, unless the conditions which I have formulated above were present.

In those cases in which antisiphilitic treatment fails and the tumour cannot be removed by the surgeon, all that can be done is to relieve the symptoms. In many cases, the pain in the back

is excruciating and the lightning (root) pains very severe. In these cases, the physician should not hesitate to administer as much morphia subcutaneously as is required to give relief. In such conditions, it is cruel to withhold morphia, as is sometimes done, lest the patient should contract the morphia habit! Whether the patient acquires the morphia habit or not, it is the duty of the physician to relieve his sufferings and to make the short period of life which remains to him as comfortable and pleasant as possible.

LECTURE XXXV

TRAUMATIC LESIONS OF THE SPINAL CORD; CONCUSSION OF THE SPINE AND SPINAL CORD, MORE ESPECIALLY IN THEIR RELATIONSHIP WITH RAILWAY ACCIDENTS AND INJURIES

TO-DAY, Gentlemen, I propose to direct your attention to concussion of the spinal cord and the lesions of the spinal cord which result from traumatic violence. The subject is a very important one. Before you have been many years in practice, most of you will probably meet with some cases in which nervous symptoms are developed after railway accidents and injuries. It is very desirable that you should enter practice with a clear understanding as to the nature and significance of these cases. We know much more about the subject now than we did a few years ago.¹

Meaning of Terms.—The first question which we have to consider is naturally one of definition. What is meant, or what should be meant, by the terms ‘concussion of the spine’ and ‘concussion of the spinal cord’? It is unnecessary to say that the term ‘concussion of the spine’ is of much wider significance than the term ‘concussion of the spinal cord’; and yet in the estimation of the public and of the lawyers they are commonly regarded as the same thing. Indeed, the popular idea of concussion of the spine and concussion of the spinal cord is even more indefinite than this statement would imply. A man who receives a general shake in a railway collision, and who may not have received any direct injury to the back or spine, very frequently subsequently suffers from a train of distressing nervous symptoms, amongst which pain in the back is often prominent.

¹ I am glad to see that in the edition of Erichsen’s great work on Surgery which has just been published, the sections on concussion of the spinal cord have been re-written, and that many of the opinions which were formerly advocated have been entirely abandoned.

Such a patient is popularly said to be suffering from concussion of the spine; the public and the lawyers, and in some cases even the doctors too, conclude that the symptoms which he manifests are due to concussion of the spinal cord, and are apt to be followed by grave organic disease of the spinal cord. These conclusions are, I hold, in most cases altogether unwarranted. In the great majority of cases of this description, there is, in my opinion, no sufficient evidence to show that the patient did, at the time of the accident, receive a concussion of the spinal cord, or that the symptoms which subsequently develop are the result of an organic lesion of the spinal cord, of the spinal membranes, or, indeed, of any part of the nervous apparatus.

The differences of opinion which exist in the minds of the profession as to the nature and significance of these cases are no doubt partly due to the fact that two separate and distinct meanings may be attached to the term 'concussion' and that the two separate and distinct things which these two meanings imply are frequently confounded one with the other. In one sense, the term 'concussion' is used to mean the mode of injury or violence. Every blow on the back and every sudden jar or wrench of the spine, the result of external violence, may perhaps in this sense be termed a concussion of the spine. In another sense, the term 'concussion' is used to mean the effect, the functional or structural change, which is produced, or which is supposed to be produced (but this is a very different thing) in the spinal cord by such injuries.

Spinal sprain. ('Concussion of the Spine').—The following case is an illustration of what may, I think, be appropriately termed a very severe sprain of the back, but which some people would call a severe concussion of the spine, without any concussion of the cord:—

Case I. Sprain of back.—A gentleman, aged 30, was riding a steeplechase in March 1893, the ground being very dry and hard. While going at great speed, his horse fell after jumping a fence, and the rider was thrown forwards with great force, the right side of the face and right shoulder striking the hard ground. After picking himself up, for he was not rendered insensible, he immediately experienced severe pain in the chest and back at the level of the mid-sternum and interscapular region, great difficulty in breathing, and a feeling of suffocation. The pain and difficulty in breathing were greatly increased by the weight of the head, shoulders, and arms, and

were to some extent relieved by allowing the head to fall forwards, and by having the shoulders and arms supported on each side. There was no pain, numbness, or loss of power in the arms or legs, and in fact no spinal (nerve) symptoms. The patient managed to get home, a distance of some thirty miles by train, with very great difficulty; the pain was so intense that at one of the intermediate stations he had to send for a doctor and have a hypodermic injection of morphine. On arriving at his destination he walked home, a distance of some two hundred yards. On getting home he immediately went to bed. On the evening of the accident the urine had to be drawn off with the catheter. The retention of urine was thought by the medical men who were in attendance to be due to the pain and general shock from which the patient was suffering and to the constrained position in which he was obliged to lie (the least movement causing intense suffering) rather than to any definite cord injury; for, on the following day, the bladder acted naturally. There was never any numbness, tingling, or loss of power in the legs. For several days, the patient suffered intense pain in the back and chest. For two days and nights, it was necessary to support the arms above the head, as the weight of the shoulders and arms pressing against the chest caused great aggravation of the suffering. For several days, he was kept under the influence of morphine. There was no fracture or dislocation of the spine; there was never any local discoloration, but there was marked tenderness on pressure over the spines of the third, fourth, and fifth dorsal vertebræ. The patient, who is by nature extremely active and energetic, was with great difficulty persuaded to remain in bed for five weeks after the accident. During the latter part of this period and for three weeks after he was allowed to move about, the spinal column was enclosed in a poroplastic jacket. In the course of two months, the patient was able to follow his profession, which involves much active outdoor exercise. Five months after the injury, he still felt some deep-seated pain in the back on bending the spine and on violent muscular effort. From first to last, the general health was good. During the whole course of the case there were absolutely no nerve-symptoms whatever. Six months after the accident he looked the picture of health and was actively following his arduous professional duties.

In this case the occurrence of retention of urine may by some be considered to have been the result of concussion of the spinal cord; if so, it was certainly the only result; but this supposition is, I think, quite unnecessary. For my present purpose, the temporary retention of urine is of no significance. The point I wish to emphasise is that in this very severe case of concussion of the spine (and, as I shall presently show, the same holds good in a large group of cases in which there is true concussion of the spinal cord),

the patient did not manifest any of the symptoms which follow (not, be it observed, in rare and exceptional cases, but in a large proportion of cases) railway accidents and injuries, in which the degree of violence which is directly applied to the spinal column is infinitely less than in the case which I have just related.

It is, of course, impossible to draw general conclusions from individual cases; but I maintain *firstly*, that cases of concussion of the spine similar in kind to, though perhaps seldom so severe in degree as, that which was present in the case which I have just related, in which none of the symptoms indicative of 'railway spine' are subsequently developed, are often met with in ordinary practice; and *secondly*, that such a severe concussion of the spine as was present in this case would, if the injury had been due to a railway collision, almost certainly have been followed by the usual train of nervous symptoms, which, for the sake of brevity, I may describe as indicative of 'railway spine.'

From this statement you will see that I agree with Mr. Herbert Page (who has done more than any one else to clear up the true nature of this most difficult subject) and others who think that in some 'railway cases' the pain and stiffness in the back are due to injury—sprains, twists, etc.—of the external parts (muscles, ligaments, bones and joints of the spinal column) and not to a lesion of the cord or its membranes. In 'railway cases,' these symptoms (pain and stiffness in the back) are rarely, I believe, indicative of inflammation within the spinal canal, that is, of meningitis or myelitis. I need not, however, dwell on this point; I shall return to it presently. I repeat that if we are to come to an agreement as to the nature of these cases, we must draw a sharp line of distinction between 'concussion of the spine' and 'concussion of the spinal cord.'

Definition of the term Concussion of the Spinal Cord.—It is not less important to come to a definite understanding as to the meaning of the term 'concussion of the spinal cord.'

The term 'concussion of the spinal cord' should, strictly speaking, be limited to those cases in which undoubted symptoms indicative of derangement of the functions of the spinal cord are directly due to a shock communicated to the cord as the result of external violence, and in which there is no obvious naked-eye lesion in the bones, membranes or cord itself to account for the condition.

I do not deny that in some cases of concussion of the spinal cord properly so called, in which the symptoms (loss of power and numbness in the lower extremities, retention of urine, etc.) last for several days or weeks, but in which complete recovery ultimately takes place, there may perhaps have been capillary hæmorrhages into the tissues of the cord and especially into the grey matter. In the comparatively speaking rare cases in which an injury to the back is, after an interval of time, followed by undoubted organic disease (sclerosis, transverse myelitis, etc.) the occurrence of minute capillary hæmorrhages, followed after an interval by inflammatory or sclerotic changes, is, I think, the most probable mode of causation.

It may of course be argued, if we accept the definition which I have just laid down, that it is impossible, unless the patient dies, to prove whether the case is one of concussion or of contusion—whether the symptoms are the result of concussion without an obvious naked-eye lesion (but with perhaps minute capillary hæmorrhages), or of contusion with an obvious naked-eye lesion such as extravasation of blood. This of course must be admitted.

Further, it may be argued that, since patients who have been injured in railway collisions rarely die, and since it is therefore impossible to apply the post-mortem test in the vast majority of railway cases, it is impossible to say whether patients who are suffering from 'railway spine' are suffering from the effects of concussion of the cord or not. This objection would of course hold good if it were admitted that the nervous symptoms which are present in cases of 'railway spine' are of spinal origin. But this is just one of the points in dispute.

But, granting for the sake of argument that this objection is valid, it may be stated that the only method of arriving at a satisfactory conclusion as to the nature and significance of the nervous symptoms which are present in cases of 'railway spine,' and which it may be alleged are the result of concussion of the cord and of subsequent organic disease produced in the cord or its membranes by the concussion, is to contrast the character, progress, and course of the nervous symptoms which are present in cases of 'railway spine,' with the character, progress, and course of the nervous symptoms which result from undoubted traumatic injuries of the spinal cord, undoubted concussion of the spinal

cord, and undoubted disease of the spinal cord in ordinary everyday practice, and to see whether in the two classes of cases they correspond or differ.

Now, the spinal lesions and diseases which result from traumatic violence in ordinary practice may be arranged in the following groups:—

I. Cases in which the spinal bones are fractured or dislocated, and in which the cord is contused or compressed by the bone lesion.

Every one will, I suppose, agree that these cases should not be included under the term 'concussion of the spinal cord,' and that cases of this kind, though common enough in ordinary practice, very rarely indeed result from railway accidents and injuries. I need not occupy time in detailing the nature of the nervous symptoms which are characteristic of these cases. We have already studied them in detail (see myelitis, compression myelitis, spinal meningitis). The significance of the symptoms in cases of this kind is obvious.

II. Cases in which, after a blow or fall on the back, a twist of the spine or a violent concussion of the spine due to a fall on the feet from a height, grave and persisting (not merely temporary and evanescent), but not necessarily permanent symptoms indicative of interruption of the function of the cord immediately follow upon the receipt of the injury, but in which there is no evidence of fracture or dislocation of the vertebral column.

In these cases, the symptoms practically correspond to those due to fracture or dislocation of the vertebræ, in fact in many of these cases there is a fracture or dislocation of the vertebræ which is only detected after death; in others, as Mr. Thorburn has shown, there is an extravasation of blood into the substance or on to the surface of the cord, but no fracture or dislocation. In some of the cases included in this group, the cord is crushed and contused by a sudden twist of the spine in the more mobile cervical region. But whatever the exact nature of the lesion, there is no mistaking the significance of the symptoms. They are identical with the symptoms which characterise acute myelitis, acute spinal meningitis, or acute compression of the spinal cord due to disease (loss of motion and sensation below the seat of the injury, paralysis of the bladder and rectum, bed-sores, elevation of temperature, priapism, etc., in

the case of acute transverse myelitis; pains, spasms, hyperæsthesia limited to nerve districts corresponding to the seat of the lesion, followed by sensory and motor paralysis in the case of meningitis, etc.). In cases of this kind (myelitis and spinal meningitis due to disease and traumatic injury) cerebral symptoms are, as a rule, completely absent unless the head has been injured, and the symptoms of general nervousness, which are so prominent in the great majority of cases of so-called 'concussion of the spine' the result of railway accidents, are rarely observed.

No one with the smallest pretence to medical knowledge could have any difficulty in concluding that in such cases the symptoms are indicative of a grave lesion of the spinal cord or its membranes. These cases should not, in my opinion, be included under the term 'concussion of the spinal cord.' Cases of this kind very rarely indeed result from railway accidents and injuries, and in any case in which symptoms characteristic of this group of cases did result from a railway accident there would be no difficulty as to the diagnosis. These are not the cases which give rise to differences of opinion in Courts of Law.

In illustration of this group I may quote the following case:—

Case II. Grave spinal symptoms without evidence of fracture or dislocation of the vertebrae.—On September 29th, 1892, Mrs. H., aged 62, being tired with a long walk, sat down to rest on a wall, which was low on the roadside but high on the opposite side, while her friends went to get a cab to take her home. While in this position, she fell over the high side of the wall (in consequence, she says, of a sudden attack of giddiness), a distance of about 8 feet, striking her head and neck against some stones. She was rendered unconscious. When picked up some little time afterwards, both arms and both legs were found to be completely paralysed. She was admitted into one of the surgical wards of the Edinburgh Royal Infirmary under the care of Dr. Peter Maclaren on October 5th, 1892; and was transferred to my care on November 7th. At this date, all four limbs were still paralysed; the muscles of both hands and forearms were markedly atrophied, and presented the usual alterations characteristic of the reaction of degeneration; the knee-jerks were greatly exaggerated, and ankle-clonus was present in each leg; the bladder was completely paralysed; bed-sores had developed; the lower limbs and trunk up to the level of the sixth rib were almost completely anæsthetic; there was also complete anæsthesia in the right arm and some

anæsthesia (the exact distribution of which it is unnecessary to detail) in the left arm. The spinal column presented no evidence of fracture or dislocation. At the time of her admission to hospital, the patient complained of a dull pain in the upper dorsal region, but there was never any fever, tenderness on pressure, localised swelling, or evidence of displacement of the vertebræ. In the course of six weeks, under the administration of iodide of potassium and hydrobromic acid, considerable improvement took place. The anæsthesia lessened; motor power was in some degree regained in the legs and in the right arm (in the shoulder and elbow); and slight movement could be effected in the right shoulder-joint. At this date, the patient insisted, contrary to advice, on leaving the hospital; she was dissatisfied because she was not allowed the free quantity of whisky to which she had been accustomed. I had no opportunity of seeing her again, but I learned that she died about the middle of July.

While believing that hæmorrhage into the substance of the cord or into the spinal canal is the most probable cause of suddenly or rapidly developed paraplegia in the traumatic cases in which there is no lesion of the bony canal, it must be admitted that such extravasations are with difficulty produced; the spinal cord is, in fact, one of the most securely protected organs in the body. The most striking illustration of this fact with which I am acquainted, came under my observation while I was acting as pathologist to the Edinburgh Royal Infirmary some years ago; the leading details of the case are as follows:—

Case III. Very severe injury of the abdominal viscera and muscles of the back due to direct violence; complete absence of spinal lesion.—J. F., aged 30, was jammed between a railway train and the platform on February 18th, 1884, and was admitted to the Edinburgh Royal Infirmary in a state of profound collapse; he died some hours afterwards. On post-mortem examination, the peritoneal cavity contained six ounces of fluid blood; *the duodenum was completely torn across in two places*,—one of the tears extended deep into the mesentery, which was infiltrated with blood; the divided ends of the intestine, which were ragged-looking and swollen, were firmly contracted, and no fæcal matter was present in the cavity of the abdomen; there was very slight commencing peritonitis; all the other abdominal organs were healthy; *each psoas muscle was ruptured*, the right being completely torn across, the lower ruptured end of each psoas muscle was firmly contracted, and formed a ball just within the brim of the pelvis; *the subcutaneous tissues and muscles of the back were extensively infiltrated with blood*, but there was no external evidence of bruising; *the spine was uninjured; the spinal cord, its membranes and nerves, were absolutely healthy.*

III. Cases in which, after a blow or fall on the back, a twist of the spine, or a fall on the feet from a height, typical and characteristic symptoms indicative of undoubted disease of the spinal cord or its membranes are subsequently developed.

In some cases of this kind, there are indications of derangement of the functions of the cord immediately after the accident, but in others there are no immediate spinal nervous symptoms, the cord symptoms being only developed after an interval—it may be after a considerable interval—of time.

Cases of this kind, in which the subsequent disease can be clearly or with great probability attributed to the injury, are, in my experience, uncommon, but I have met with several which resulted from ordinary injuries. I have never seen a single case which resulted from a railway accident or injury, but such cases do, no doubt, occur. They are, however, quite exceptional and extremely rare. Let me briefly detail the facts of two cases of this kind which resulted from ordinary injuries:—

Case IV. Diving accident: late development of spinal symptoms.—A strong and muscular lad, aged 14, dived into a shallow pool at the beginning of August 1892, striking his head against the sand, the depth of the water being about 2 feet. His head was not injured, but he gave his neck a tremendous twist. On getting up he felt somewhat stunned, but was able to dress himself and walk home, a distance of a hundred yards; he experienced some tingling in the fingers of the left hand, but this symptom was not marked; he complained more of pain in the shoulders than of anything else. In consequence of the pain in the shoulders he was kept in bed until the end of August, when his father, who was travelling on the Continent, returned to this country. He was then removed from Wales to his home in Aberdeen. There had never been any swelling of the neck, or any projection of the bones, but the head was kept stiff and inclined towards the left side, apparently because of pain. At the beginning of October he seemed to have recovered so completely that he was allowed to attend private classes. After being at school for a week or ten days, while bending the spine, he felt a sudden pain in the left shoulder, followed by numbness and loss of power in the arms, the left being more affected than the right. The paralysis in the arms quickly increased, and the legs soon became affected. In the course of a short time the muscles of the forearms and hands became markedly atrophied. At this time the neck was freely movable and free from pain; there was no local tenderness or swelling; no fever, no alteration in the pulse rate; in short, no constitutional disturbance. On October 12th, when I was consulted by letter, the symptoms were increasing in severity.

At this date there was complete loss of motor power in the left arm and hand, less complete but marked paralysis in the right arm and hand and in both lower extremities. Sensation in the upper and lower extremities and in the trunk was markedly impaired. The paralysed parts were rigid, and violent spasmodic twitchings, which were so severe as almost to resemble tetanic spasms, occurred both in the arms and legs, but especially in the arms; the rigidity and spasms were worse in the morning and after waking from sleep. The deep reflexes, both in the upper and lower extremities, were extremely exaggerated. The bladder was not paralysed, but there was obstinate constipation. I advised that the patient should be kept absolutely quiet in bed, with the neck fixed and supported by a pillow; that the actual cautery should be freely applied on each side of the lower cervical and upper dorsal spines, and that iodide of potassium and hydrobromic acid should be given internally. My friend Dr. Blaikie Smith, who was in attendance on the patient, tells me that the application of the actual cautery was soon followed by very marked improvement, which has continued uninterruptedly up to the present time. The patient is now quite well; motor power has been completely regained, both in the arms and legs; there is still, however, slight exaggeration of the deep reflexes in the lower extremities. His father, who is a distinguished member of the profession, in writing to me on July 26th, 1893, said: 'I shall say, to my dying day, that the actual cautery saved him.'

This case should, perhaps, be included in the preceding (second) group, for there were distinct and definite symptoms indicative of spinal injury immediately after the accident. I have included it in the third group, because the chief symptoms did not develop until two months after the original injury.

Opinions may differ as to the exact nature of the lesion which developed two months after the injury. My own view is that it was situated on the surface rather than in the substance of the cord, and that it was probably a sudden extravasation of blood or inflammatory effusion which had implicated the nerve roots in the lower cervical and upper dorsal region, and had pressed upon and interrupted the functions of the spinal cord. But be this as it may, there could, of course, be no doubt as to the serious and organic nature of the condition.

In the following case there were no symptoms indicative of a lesion of the cord at the time of the accident; the spinal symptoms did not develop until six weeks after the accident. The case is as follows:—

Case V. Fall on left side: late development of transverse myelitis.—

J. U., aged 41, a strongly-built and robust-looking man, consulted me at the Edinburgh Royal Infirmary on July 8th, 1893, on account of paraplegia due, he stated, to a fall received eighteen years previously. He gave me the following account of the accident and of his illness. While pole vaulting, his pole broke, and he fell to the ground from a height of 9 ft. 3 in., alighting on his left hand and knee. Immediately after the accident he felt slight pain in the small of the back, but no numbness or weakness in the legs; in short, no spinal symptoms. For three weeks he was able to walk about with a limp, suffering pain in the left knee, and some slight dull pain in the left lumbar region. At the end of three weeks he felt, he says, quite well; the pain in the back had disappeared; there was no weakness or numbness in the legs, and there was no difficulty in making water. For six weeks after the accident he followed his occupation, which was that of a gasfitter. Previous to the accident he had been a very healthy man, was married, and had never had syphilis. One Wednesday, six weeks after his fall, when standing on the top of a high ladder mending a gas bracket, he felt as if he had been suddenly struck behind the right knee. He immediately came down the ladder, feeling shaky all over. He continued to follow his employment until the following Saturday, feeling weak and shaky in the legs, and complaining of 'a sort of dull pain in the back.' The weakness could not, however, have amounted to much, for on the Saturday morning he went to his work at 6 o'clock, walking a distance of four miles. At 10.30 he left off work and walked the four miles home again, but with the greatest difficulty. On getting home he went to bed and sent for a medical man. On Monday morning he attempted to rise and go to work, found that he was unable to stand and fell on the floor, in consequence, he says, of loss of power in the legs. After this date he rapidly got worse; in the course of a few days the legs became completely paralysed, complete anæsthesia developed in the lower extremities and in the trunk up to the level of the waist, the bladder became paralysed, and bed-sores developed. He was entirely confined to bed for nine months. During this time his temper was, he says, irritable, but he did not manifest any of the nervous symptoms such as one sees in cases of railway spine. At the end of a year he began to regain a little power in the lower extremities, and in the course of time was able to stand and then to walk. The improvement in motor power very slowly continued during the first eight years after the accident. At the end of ten years sensation was, he says, completely restored. At the end of thirteen years he regained power over the bladder; up to that time it had been entirely paralysed, but the urine had never been ammoniacal or fœtid. When I saw him, eighteen years after the accident, he was able to walk, but only with great effort and difficulty; a distance of a hundred yards seriously exhausted him. The gait was typically spastic in character, the left leg being

somewhat worse than the right; the muscles of the lower extremities were well nourished; the knee-jerks were greatly exaggerated, and ankle-clonus was present in both legs; sensibility of all kinds was, practically speaking, perfect both in the lower extremities and in the trunk; and there was no paralysis of the bladder. There were several scars on the back, some due to bed-sores, others to the actual cautery, which had been several times applied over the lumbar and sacral regions. There were also several cicatrices on the lower extremities, the results of trophic ulcers which had formed from time to time during the first year of his illness.

It may of course be said that in this case the transverse myelitis, which was obviously the cause of the symptoms, was not due to the accident at all; that its occurrence six weeks after the original injury was a mere coincidence. But with this view I am not disposed to agree. It seems to me that the myelitis was probably the result of an injury to the cord, perhaps a small extravasation of blood into the grey matter produced at the time of the accident. The facts that the patient was up to the time of the accident an unusually strong, healthy, and robust man, that immediately after the accident he felt a dull pain in the back, and that this pain continued for the three weeks after the accident, seem to suggest a direct connection between the accident and the subsequent myelitis.

In addition to these cases, both of which are of great interest, I have of course met with other cases in which a traumatic injury, blow or fall on the back, or a fall from a height on the feet or nates, appeared to be the direct cause—the exciting cause at all events—of an organic lesion of the spinal cord which followed it.

Chronic Spinal Meningitis.—I have said nothing, you will observe, of chronic spinal meningitis. In my experience, chronic spinal meningitis due to traumatic injury (blows or falls on the back)—I do not, of course, include cases of pachymeningitis due to fracture of the spine or to Pott's disease following an injury, or to cases of syphilitic (gummatous) meningitis—is an extremely rare condition. In fact, I may go further than this, and say that chronic spinal meningitis—if we exclude (a) pachymeningitis externa due to bone disease, (b) pachymeningitis cervicalis hypertrophica, (c) leptomeningitis and associated pachymeningitis due to syphilis, (d) leptomeningitis of the localised form associated with definite and distinct lesions of the cord

such as locomotor ataxia, and (e) chronic leptomeningitis the result of a definite and distinct attack of severe acute leptomeningitis spinalis, in all of which conditions the nature of the case is usually clear enough—is exceedingly rare.

Further, I would point out that chronic spinal meningitis (leptomeningitis) when it does occur is usually associated with myelitis, and that it is a grave lesion; that in most cases its course is progressive; and that, although in its earlier stages the diagnosis is often very difficult, in its later periods the symptoms are clearly and unmistakably indicative of a grave organic lesion of the cord or of the nerve roots which happen to be implicated.

Late Occurrence of Gross Lesions.—In my experience, in ordinary private practice and in persons who were previously healthy, undoubted and unmistakable lesions of the spinal cord (such as transverse or disseminated myelitis, locomotor ataxia, spastic paraplegia, disseminated sclerosis, and especially chronic spinal meningitis) can only in rare instances be attributed to traumatic injuries of the back. I have never seen an undoubted case of any of these cord lesions follow a railway accident or injury.

These facts, if they be facts, are of the greatest significance. Severe injuries to the back, immediately followed by definite and distinct symptoms indicative of concussion of the spinal cord, are, as I shall presently show, of common occurrence, but *provided that they are unattended with fracture or dislocation of the vertebrae* it is only in exceptional and rare cases that they are followed by organic disease of the spinal cord or its membranes.

Transverse myelitis, spastic paraplegia, locomotor ataxia, and progressive muscular atrophy are common diseases, but it is only in a small proportion of cases that they can, with any degree of probability, be directly attributed to traumatic injury.

It is a remarkable fact that these undoubted cord lesions are very rarely, if ever, met with in persons who have been injured in railway accidents. Some of the diseases which I have just enumerated are so common that, on the mere theory of chances, cases must occasionally occur in persons who have been injured in railway collisions. As I have already said, I have never myself met with any case of this kind, but it is certain that some of them must occasionally occur. The wonder is not that they should, if they do, occasionally occur, but that they should so very rarely

occur. The mere fact that such a disease as locomotor ataxia develops after an injury to the back does not, of course, show that it was due to the back injury. The association may be merely accidental. The special facts and circumstances of each case must be carefully and judicially considered before coming to the conclusion that the disease is due to the injury.

That the receipt of a local injury often directs attention to symptoms or even physical alterations in the bodily conformation, which have been slowly and imperceptibly developing for weeks, months, or even years, and that in cases of this description the patient very naturally concludes that the injury was the cause of the affection which he for the first time perceives, is of course well known. In every case, therefore, in which a blow on the back or other traumatic injury seems to have been the cause of organic disease of the spinal cord, it is of the greatest importance to inquire carefully into the history, and to make quite certain that the disease was not already present before the receipt of the injury. When the injury or accident seems to have been the starting-point of the affection, it is also of importance to ascertain whether there was not some hereditary or acquired tendency to the special disease from which the patient is suffering, which, being previously latent, was, as it were, aroused into activity by the injury. It is well known, for instance, that in persons who have suffered from syphilis, an accident, an injury, mental worry, or anything else which seriously impairs and depresses the general health, is often followed by an outburst of tertiary symptoms which previously had been entirely absent. So, too, when there is a strong hereditary tendency to cancer, a local injury is often the exciting cause of the disease; it is also extremely common to find in cases of scrofulous tumour of the cerebellum, a history of local injury (a blow or fall on the back of the head). While I am quite willing to admit that all of the lesions included in the three groups enumerated above may perhaps in rare and exceptional cases result from an injury received in a railway collision, I maintain that such a mode of production is so exceptional that it constitutes no argument whatever for the belief (on the contrary, the extreme rarity of its occurrence is, I maintain, a very strong argument in opposition to the belief) that the nervous symptoms which are such a com-

mon result of railway accidents and injuries are due to organic disease of the cord or its membranes.

IV. Cases in which after direct injury, such as a fall of coal on the back, symptoms indicative of concussion of the spinal cord (temporary paralysis of the bladder, temporary numbness, and loss of power in the legs) occur.

In ordinary civil practice, concussion of the spinal cord (temporary disturbance of the functions of the spinal cord), strictly analogous to the more common concussion of the brain, is a rare condition. With the object of showing that the nervous symptoms which follow railway accidents and injuries do not depend upon concussion of the spinal cord, my friend Mr. Frederick Page makes use of the following argument:—‘ If forty people were injured in a railway collision, the probability is that thirty-five of them would subsequently suffer, or be said to suffer, from concussion of the spinal cord ; but if you were to search the records of any large surgical hospital for a period of ten years, you would probably not find ten cases entered in the books as cases of concussion of the spinal cord.’

In colliers, concussion of the spinal cord seems to be a not infrequent result of falls of coal on the back. So far as I know, this fact is not generally known to the profession ; it is one of the greatest importance in relationship to the concussion of the cord which is supposed to result from railway accidents and injuries.

Colliers are perhaps exposed more than any other class of the community to direct injury to the back in consequence of falls of coal and ‘ roof.’

In some cases (fortunately they constitute a small proportion of the whole) the spine is fractured or dislocated—usually, I am informed, as the result of a large and heavy mass of coal or ‘ roof’ falling on the upper part of the spinal column ; the head and upper part of the spinal column are suddenly and forcibly depressed ; the back seems usually to give way in the lower dorsal region. The characteristic symptoms of transverse myelitis or acute meningitis and death, or permanent paralysis, result.

In other cases (and I am informed that they frequently occur) a fall of coal or stone on the back produces temporary paralysis of the bladder, requiring the use of the catheter, and temporary numbness or loss of power in the legs. The paralysis

of the bladder not infrequently persists for several days, or even, in exceptional cases, for some weeks. Obviously, therefore, it is not merely the result of general shock; in other words, it is not analogous to the retention which so frequently occurs after fractures, surgical operations, and in fact almost every kind of severe injury. The paralysis of the bladder and the loss of power and numbness in the legs (which, according to some of the authorities I have consulted, are more common, but according to others less common, than the paralysis of the bladder) are evidently, in some cases at least, the direct result of a sudden derangement of the functions of the spinal cord produced by the fall of coal or stone on the back.

There can, I think, be no doubt whatever that in some of these cases the spinal symptoms are due to concussion of the spinal cord, properly so called.

Now, if concussion of the spinal cord is a common cause of spinal meningitis, meningo-myelitis, and organic disease of the spinal cord, we should naturally expect that colliers, whose cords have been concussed in the manner which I have just described, would frequently suffer from these cord lesions. But, as a matter of fact, they very rarely, if ever, do so.

And, again, if the symptoms of 'railway spine' are the result of concussion of the cord and of organic disease of the spinal cord and its membranes, due to such concussion, colliers ought to manifest the train of symptoms which characterise cases of 'railway spine.' But as a matter of fact they rarely, if ever, do so.

I have weighty evidence in support of all these propositions. I first learned the frequency with which concussion of the cord is produced by falls of coal on the back from my friend Dr. Drysdale of Dunfermline. He writes me as follows:—

I have during the last eighteen years frequently met with cases in which the bladder has been paralysed for two or three days, sometimes longer, after bruising of the back from falls of coal or stones. Indeed, it has been my usual habit when I have been called to a pit accident of this kind to put a catheter in my pocket. I have also frequently seen temporary loss of power in the legs as the result of these injuries, but as regards this it must be remembered that it is the usual habit of the average collier, when he meets with an injury, to declare that he has lost 'all power,' and to more or less exaggerate his condition. I have never seen permanent disease of the spinal cord except in those

cases in which there was undoubted injury to the spinal column. In the ordinary cases of simple bruising from falls of coal or stones where there is paralysis of the bladder lasting for a few days with slight loss of power in the legs quickly passing off, you get complete recovery in a few weeks. I cannot recall any such case which was followed by permanent disease.

I have also consulted Dr. Nasmyth, now Medical Officer of Health for the County of Fife, who for many years had medical charge of large collieries at Cowdenbeath; Dr. Lennox of Hamilton, who has had fifty years' experience of colliery practice; Dr. Crawford of Hamilton, who has also had a very large experience; Dr. Anderson of Seaton Delaval, and Dr. S. W. Broadbent of Castle Eden, who have probably had a larger colliery experience in the great coalfields of Northumberland and Durham than any other living men; and Dr. Thomas and Dr. Warburton of the Rhondda Valley, who have had a vast experience in the coalfield of South Wales. All of these gentlemen, whose opinion may be taken as authoritative, are agreed as to the frequency of temporary paralysis of the bladder, and most of them are agreed as to the frequency of temporary loss of power and numbness in the legs after these accidents. No one of them has ever seen organic disease of the spinal cord result from a fall of coal on the back, except in those cases in which the spinal column was severely injured. Only one of them has seen, and that only in a single instance, symptoms characteristic of 'railway spine' result from a fall of coal on the back, or from the concussion of the spinal cord which this injury so frequently seems to produce.

I shall not be surprised to hear that in some instances organic disease of the spinal cord does result from falls of coal or stone on the back; indeed, I am astonished that none of the gentlemen whom I have consulted have met with any case of this kind. But even granting—as I am quite prepared to grant—that if organic disease of the spinal cord does occasionally follow concussion of the cord (I mean, of course, concussion pure and simple) due to a fall of coal on the back, the large body of evidence which I have collected conclusively, I think, shows—

Firstly, that in colliers falls of coal or stone on the back frequently produce concussion of the spinal cord.

Secondly, that the symptoms indicative of such concussion are in the vast majority of cases merely temporary.

Thirdly, that colliers who have suffered from concussion of the spinal cord (I mean, of course, concussion pure and simple) very rarely subsequently suffer from organic disease of the spinal cord or its membranes.

Fourthly, that they very rarely indeed manifest the train of nervous symptoms which so frequently occur after railway accidents and injuries.

The two last conclusions may in the future be of no small importance, quite irrespective of the subject of railway accidents and injuries with which we are at present immediately concerned. The tendency of modern legislation is to compensate *employés* who have been injured, provided that the accident is not the result of their own carelessness or error. Now I venture to predict that if the Legislature should enact that colliers are entitled to compensation for the injuries so received, nervous symptoms will in the future be found to result much more frequently from falls of coal and stone on the back than is at present the case.

V. Cases of so-called 'railway spine,' or cases of traumatic neurosis due to railway accidents and injuries.

Of the numerous and diverse symptoms which these cases present many are clearly due to derangement of the functions of the brain rather than of the spinal cord. By the public and the lawyers these cases are not infrequently termed cases of 'concussion of the spinal cord' or 'concussion of the spine'; scientifically and medically they are best, I think, termed cases of traumatic neurosis due to railway accidents and injuries.

Before describing the symptoms in detail I must distinctly state that I refer to the ordinary common typical cases in which nervous symptoms are developed after railway accidents and injuries, and not to cases which should be included in the first, second, or third of the preceding groups. Nor do I refer to cases in which a localised paralysis follows an injury of one or more of the peripheral nerves; nor to cases in which a peripheral neuritis has resulted from a definite local injury. These peripheral nerve lesions, with their characteristic symptoms, no doubt occasionally occur after railway accidents and injuries, but, like the unmistakable organic lesions of the spinal cord

included in the first, second, and third groups, they are, so far as my information enables me to judge, very rare.

The traumatic neuroses are extremely common after railway accidents and injuries, while, after ordinary injuries, they are, comparatively speaking, less frequent.

Symptoms of 'Railway Spine.'—In my experience the following are some of the most common and characteristic symptoms which patients who have been injured in railway accidents complain of (I refer especially to cases in which the symptoms appear to be genuine, and in which the patients are not intentionally exaggerating or malingering):—

General weakness, prostration, and exhaustion.

Extreme nervousness; loss of nerve tone; 'loss of nerve.'

Sleeplessness; broken and disturbed sleep; unpleasant and frightful dreams—the patient often starts out of a terrifying dream thinking that he is in a railway collision or that he is falling over a precipice.

Confusion of thought.

Failure of memory.

Diminished emotional control.

Inability to concentrate the thoughts; inability to calculate; inability to read; inability to occupy the attention with business matters; total business inaptitude.

Uneasy sensations in the head, rarely amounting to distinct or severe headache; a feeling of weight, fulness, oppression, throbbing, tightness, or constriction in the head; or of pressure on the vertex.

Temporary giddiness.

Irritability of temper.

Change of disposition; a tendency to be upset and worried by trifles which before the accident would have given the patient no concern.

Black specks before the eyes (*muscae volitantes*).

Weakness of sight. This never in my experience amounts to continuous loss of vision; in the cases which have come under my own notice the failure of sight was merely temporary. In many cases when the patient attempts to read, letters, words, and lines are said to run together, and the vision becomes confused or fails.

Photophobia.

Noises in the head or ears ; hypersensitiveness to noise.

Palpitation of the heart.

Pain in the back, in many cases increased on movement of the spinal column and on muscular effort.

Difficulty in walking ; inability to walk ; a feeling of extreme exhaustion and fatigue after walking a short distance ; loss of power in the legs, less frequently in the arms.

Numbness, deadness, uneasy sensations, but rarely sharp, shooting, lancinating pains, in the legs or arms ; pain in the legs on movement ; startings and jumpings in the legs or trunk on falling to sleep.

Loss of sexual desire and sexual power.

Constipation.

Irritability of the bladder ; in some cases slight loss of expulsive power in the bladder ; never in my experience paralysis of the sphincters or persistent retention.

Loss of appetite.

Loss of weight.

The physical condition of patients who are suffering from 'railway spine.'—The patient has in many cases a careworn, anxious, and exhausted expression. His friends and acquaintances often say that 'he is quite a different man' ; that 'he is visibly changed' ; that 'he is ten years older than he was before the accident,' etc.

Tenderness rarely limited to one point, but generally diffused over the whole spine, or present in several distinct positions (in the cervical, dorsal, and lumbar regions), can often be elicited on pressure over, or percussion of, the vertebral spines.

In many cases the skin over the vertebral spines or down the whole of the back is extremely sensitive to pressure (hyperæsthesia).

In some cases there is rigidity of the spine. This condition has, in my experience, only been well marked in those cases in which the back has been obviously bruised or strained at the time of the accident, and in which the spinal pain and tenderness have been developed immediately or soon after the injury. But, in all cases in which the pain in the back is prominent (I do not refer to mere hyperæsthesia of the skin) the patient may of course, in order to avoid movements which increase the pain, keep the back more or less rigid and stiff.

Tenderness on pressure over the spine and hyperæsthesia of the skin of the back are very often developed in cases in which there is no evidence that the back was directly injured, sprained, or bruised at the time of the accident. In some cases these symptoms are only slightly marked in the earlier stages of the case, and only become prominent when the general nervous symptoms which I have already described are fully developed.

The patient often walks with an uncertain, feeble, or apparently feeble gait, supporting himself in his efforts to cross the room by leaning with his hand on the table, chairs, etc. In many cases one leg is said to be weaker than the other. In such cases the weak leg may be dragged, but it is usually kept stiff, and the foot moved forward as a whole, the heel touching the ground. The toes do not drag or catch the floor, and the leg is not circumducted as in ordinary hemiplegia. There is no 'high action' or typically spastic gait.

The grasping power of one hand, as measured by the dynamometer, is often less than that of the other; but as every one knows, this is not a sign of the existence of paralysis due to organic disease, or, indeed, of paralysis at all.

The muscles are in some cases soft and flabby, in others fairly firm and well nourished. But in the cases to which I am referring there is rarely any localised atrophy and wasting.

The knee-jerk is in many cases exaggerated, sometimes normal or subnormal in degree. Well marked and typical ankle clonus is rarely, if ever, present. Provided that proper precautions are taken in conducting the examination, the knee-jerks will usually be found to be equal on the two sides. I have never seen a case in which the knee reflex could not be elicited. The plantar reflex is usually well marked, often exaggerated—a condition which is common in many neurotic and in fact in many normal conditions.

Retention of the urine due to paralysis of the detrusor, and incontinence due to paralysis of the sphincter, have never in my experience been present.

Localised patches of anaesthesia or hyperæsthesia, and perhaps more frequently hemianæsthesia or hemihyperæsthesia, are often present.

The pupils vary in size; but they are rarely unequal; they respond normally, both to light and accommodation. The visual fields are said in some cases to be contracted, often more so in

one eye than in the other; and this limitation or contraction of the fields, both for white and for colours, may be more marked at the end than at the beginning of a long perimeter examination. In other words, the characteristic alterations of the visual field, which are so common in hysteria and neurasthenia, are sometimes present. I have rarely had an opportunity of making a careful perimeter measurement of the fields, and I cannot speak from personal experience on this point.

I have never seen any definite and characteristic changes, such as papillitis or optic atrophy, in cases of 'railway spine.'

To sum up: the nervous symptoms which are met with in persons who have been injured in railway collisions are for the most part entirely subjective. Definite and distinct indications of organic disease in the brain or spinal cord are rarely present.

The mode of onset, progress, and development of the symptoms vary in different cases. In some cases, there is a definite local injury (in the form of a contusion, abrasion, superficial wound, localised swelling, etc.) to the back, head, or limbs; in others, the patient has merely received a general shake, but no local bruise or injury. The disproportion between the local injury received at the time of the accident and the severity and long continuance of the subsequent symptoms is in many cases a striking and highly characteristic feature.

Mr. Herbert Page and other authorities who have had large experience in these cases point out that the symptoms which I have just described comparatively seldom occur in railway *employés* (guards, etc.) who have been injured in railway accidents; and that passengers who have sustained severe local injuries, such as fractures of the leg, are much less liable to suffer from the general nervous symptoms which I have just detailed than persons who have been apparently much less severely hurt.

In many cases the patient states that at the time of the collision he was knocked from one side of the carriage to the other, or that he was thrown on to the floor. In some cases he is rendered unconscious, much more frequently he is merely stunned or dazed. In many cases he is able to get up immediately after the accident, feeling, perhaps, ill and faint; often he is able to go about and help those of his fellow-travellers who have been more severely injured than himself. He is frequently able to proceed to his destination; in many cases

walking some distance, feeling perhaps ill, faint, and shaken, on his way home.

After the first effects of the shock have passed off, there may be a period of comparative ease, during which the patient perhaps complains of pain and uneasiness in the back, but during which the nervous symptoms, which are subsequently developed, are not yet marked; it may be only after some weeks or even months that the nervous symptoms become fully developed. In many cases, after the first effects of the accident have passed off, the patient attempts to go to business and follow his ordinary occupation, but he soon finds that he is unequal to the effort—any attempt to do business produces a feeling of extreme fatigue, and makes his head symptoms worse.

Pathology.—The exact pathology of the cases which are included in this, the fifth group, has given rise to a great deal of discussion and debate. Many different views have been held. The two most important are as follows:—

1. That the symptoms are the result of concussion of the spinal cord, of anæmia of the cord, or of subsequent organic disease in the spinal cord and the membranes of the cord (spinal meningitis and meningo-myelitis), the result of such concussion.

2. The other view is that the symptoms are the result of a functional disturbance in the nervous system; that they are, in short, indicative of a traumatic neurosis.

By some the condition has been termed traumatic neurasthenia, by others traumatic hysteria. I hardly think that either of these terms fully and accurately describes their nature, though symptoms indicative of neurasthenia and hysteria are often, of course, present.

In my opinion there can be no question that the latter view which Mr Herbert Page has done so much to establish, is the correct one. So far as my experience and observation enable me to judge, the nervous symptoms which are developed after railway accidents and injuries are in the vast majority of cases due to functional disturbance, and not to organic disease. The nature of the symptoms, their mode of development, progress, and course are, in my opinion, altogether opposed to the view that they depend upon spinal meningitis or meningo-myelitis (the organic disease of the spinal cord or of the spinal mem-

branes to which they were at one time ascribed). I am speaking, be it remembered, of the ordinary, common, typical cases. I have already stated that in exceptional instances organic lesions of the spinal cord, brain, or peripheral nerves may result from railway accidents and injuries. But I am not at present concerned with these exceptions.

In opposition to the view that the symptoms are due to spinal meningitis or meningo-myelitis, the following facts are, I think, of importance :—

1. Chronic inflammation of the membranes of the spinal cord (chronic leptomeningitis) very rarely indeed results from injuries to the back in ordinary practice.

2. Chronic spinal meningitis is extremely difficult to diagnose, more especially in its earlier stages.

3. Chronic spinal meningitis very rarely if ever results from falls of coal on the back, though concussion of the spinal cord seems to be frequently produced by such injuries.

4. Chronic spinal meningitis, when it is actually developed, is a very grave lesion and is usually associated with myelitis (meningo-myelitis).

5. The symptoms which are present in the great majority of cases of so-called 'railway spine' are not indicative of meningo-myelitis. In cases of meningo-myelitis the motor and sensory disturbances are much more definite and are much more distinctly localised (limited and distributed) in the areas of distribution of the implicated nerve roots.

6. Further, in the rare cases in which meningo-myelitis follows traumatic injury to the back in ordinary practice cerebral symptoms are usually conspicuous by their absence.

7. In cases of so-called 'railway spine,' cerebral symptoms are prominent, in fact they constitute a large part of the clinical picture of the case. In order to account for the presence of these cerebral symptoms it has been suggested that in some cases at least the cerebral symptoms are due to inflammation of the cerebral meninges, the result of an injury to the head received at the time of the accident, and that in others an inflammation, which is first excited in the spinal membranes, makes its way upwards, and subsequently affects the membranes of the brain.

Both these views are, in my opinion, untenable. Chronic cere-

bral meningitis, like chronic spinal meningitis (if we exclude the syphilitic and the alcoholic forms, and the rare cases of chronic basal meningitis which are very occasionally met with in children and young persons) is almost unknown. Further, although it is of course perfectly true that acute (purulent or tuberculous) spinal meningitis is very often associated with acute (purulent or tuberculous) cerebral meningitis, it must be remembered that in such cases the spinal meningitis is usually secondary and subordinate to the inflammation of the cerebral meninges. Cerebral meningitis seems very rarely to result from spinal meningitis, even in acute cases, and chronic cerebral meningitis secondary to, and the result of, chronic spinal meningitis is, in my experience, a condition which is practically unknown. It is one thing to say that acute inflammation of the membranes of the spinal cord is very often associated with acute inflammation of the cerebral membranes; it is another and a totally different thing to say that chronic spinal meningitis is apt to result from traumatic violence, whether applied directly to the back or to the body as a whole, and that the inflammation in the spinal membranes which is in this way established is apt to extend to, and involve, the membranes of the brain.

8. Further, the cerebral symptoms which are present in cases of so-called 'railway spine' have never, in my experience, been suggestive of an inflammation of the membranes of the brain.

9. And, again, chronic inflammation of the cerebral meninges, like chronic spinal meningitis, is a grave and serious lesion. The course which the symptoms pursue in cases of chronic cerebral meningitis is usually quite different from the course of the cerebral symptoms in cases of 'railway spine.'

These considerations conclusively, I think, show that the nervous symptoms indicative of 'railway spine' do not depend either upon cerebral or spinal meningitis. I repeat that in the great majority of cases, there is absolutely no evidence of organic disease. It is unquestionable, I think, that in the vast majority of cases the symptoms are merely functional. The disturbance no doubt in some degree affects the whole nervous system, but the derangement is chiefly cerebral and psychical. Any changes which may be present in the spinal centres are, for the most part, secondary.

Diagnosis.—A positive diagnosis of concussion of the spinal cord in the sense in which it has been defined above (see p. 615) is necessarily somewhat hazardous. True concussion of the spinal cord is rare. It may be suspected when well-marked symptoms of deranged spinal action follow a blow on the back, a fall on the feet, or other injury in which a severe shake or jar has been communicated to the spinal column, and in which, so far as can be ascertained the spine has not been fractured or dislocated. A positive diagnosis of concussion of the spinal cord pure and simple is only justified when the spinal symptoms, which have been produced in this way, rapidly and completely disappear without leaving any permanent bad effects behind them.

In cases in which well-marked spinal symptoms follow a blow on the back or other traumatic injury, the object of the physician should be to determine:—(1) Whether the symptoms are genuine; (2) whether the symptoms are indicative of organic disease or of functional derangement; (3) if the symptoms are due to organic disease, the exact situation, extent and nature of the lesion; (4) if there are no distinct indications of organic disease, whether the symptoms are due to derangement (a) of the functions of the spinal cord, (b) of the brain, or (c) of both the spinal cord and brain.

In order to arrive at a satisfactory conclusion on these points, a thorough acquaintance with the whole subject of nervous diseases is necessary. In order to arrive at a correct diagnosis, the case must be thoroughly and carefully examined and a judicial opinion formed as to the significance of the facts just as in any ordinary case of nervous disease. In practice, it will, I think, be found convenient to classify individual cases, so far as possible, under one or other of the five great groups described above. In those cases in which there is any reason to expect exaggeration or deception—and this statement applies most forcibly to railway compensation cases—all the collateral and non-medical circumstances connected with the case, such as the whole behaviour of the patient and the amount of compensation which he is claiming, must be taken into account.

Prognosis.—In all cases in which nervous symptoms appear to be the result of traumatic injury the prognosis must, of course,

be based upon:—The character and severity of the symptoms; the nature of the lesion or functional derangement which seems to be present; the nature and severity of the injury; the way in which the symptoms developed; the individual peculiarities of each patient; and the collateral (non-medical) considerations of each case. The opinion must also, of course, be guided by the general course which the individual forms of organic lesion and functional derangement (classified in the different groups enumerated above) tend to pursue; and in judging of this course the physician must rely upon his own special experience and the published experience of other observers.

In order to come to a just conclusion as to the nature and significance of the nervous symptoms which result from railway collisions, it must be remembered:—

(1) That in the great majority of cases, the nervous symptoms are merely functional.

(2) That in some cases (and they undoubtedly constitute a very small proportion of the whole) distinct signs of organic disease are developed.

(3) That in those cases in which there are distinct signs of organic disease, the symptoms characteristic of functional derangement, general nervous shock, are often present.

While, then, on the one hand it must not be rashly and hastily supposed that because the symptoms of general nervous shock are present there is no organic disease; on the other hand, it must be remembered that, even when symptoms of organic disease are developed, many of the nervous symptoms due to general nervous shock are often present. In mixed and complicated cases of this kind, the symptoms dependent upon functional derangement may be reasonably expected to subside; while those due to organic disease may of course remain.

The future progress of cases of so-called railway spine is very variable. From what I have already said, you will readily understand that it is impossible to lay down general rules of prognosis which can be applied to individual cases; in fact, each case must be judged of on its individual merits. But speaking generally it may be said:—

(4) That in almost all cases in which the symptoms are severe and the patient is claiming compensation, the symptoms persist and get worse rather than better, so long as the anxiety

and worry attending litigation are hanging over the patient's head.

(5) That in the majority of cases in which the symptoms are merely functional, improvement commences as soon as the worry and mental anxiety attending litigation cease.

(6) That, as a rule, in cases of this description the patients ultimately get quite well.

(7) That in some cases, in which there are no signs of organic disease but in which the symptoms are merely due to functional disturbance (general nervous shock), recovery is slow and tedious.

In the great majority of 'railway cases' recovery ultimately takes place. This is more particularly the case when the patient is young, and when he was previously robust and healthy. In old, weakly and debilitated subjects, in cases in which the patients have little or no reserve of nervous energy, the severe shock to the nervous system (to speak in popular language) which they have received and the prolonged sufferings which they have gone through may have so impaired their nerve tone that they never recover their former strength and activity. But even in these—the most unpromising—cases a very considerable degree of improvement usually takes place. In some cases, the general nervous shock is so profound that although the injury has not produced any localised organic disease, the general nervous balance is so seriously deranged and damaged that the patient, for years after the accident, is an altered man, unable to enjoy the pleasure of life and unable to cope with its difficulties, and in some cases unable to carry on his ordinary business occupation.

It can hardly be doubted that a shattered condition of the nervous system materially affects the general health, lowers the resisting power of the patient, and predisposes to various diseases. It is possible, therefore, to conceive that a railway collision may actually shorten a patient's life by predisposing to a disease which at first sight may appear totally unconnected with the accident. As a matter of fact, however, cases in which such an event follows are exceedingly rare.

The course which cases of 'railway spine' go through is by no means peculiar. Exactly the same sequence of events is seen in many cases of ordinary traumatic neurosis, neurasthenia and

hysteria. We know that it is often extremely difficult to cure cases of functional (hysterical) paralysis unless the physician is firmly satisfied that there is no organic lesion. It is not, therefore, to be wondered at that in some cases of functional paralysis, the result of railway collisions, in which, be it remembered, the patient naturally concludes that the paralysis is organic (and in many cases he is confirmed in this opinion by his medical advisers), the paralysis should continue for long periods of time after the question of compensation has been finally settled.

From these statements, you will understand that I take a somewhat less hopeful view of the rapidity of recovery, and in some cases of the probability of complete recovery, than some authorities seem to do. I emphatically repeat, however, that in the great majority of cases of railway spine complete recovery does ultimately take place; that it is impossible to form a prognosis by general rules; but that each special case must be judged on its own merits.

Treatment.—The treatment of cases in which spinal symptoms follow traumatic injury must be conducted in accordance with the nature of the lesion (structural change) or functional disturbance which is supposed to be present. I need not go into details, for in previous lectures we have fully considered the treatment of the individual diseases of the spinal cord.

In treating cases of 'railway spine' in which nervous symptoms of a functional character are alone present, it is essential to remember:—

Firstly, that it is difficult, indeed in many cases impossible, to cure a functional nervous derangement unless you succeed in thoroughly convincing the patient that the symptoms are merely temporary and incurable; and that you cannot expect to succeed in persuading the patient that you can and will cure him unless you feel perfectly sure of your diagnosis.

Secondly, that the functional symptoms to which I am now referring are largely influenced by expectant attention and by the worries and anxieties attending the question of compensation and its settlement. In order, then, to cure the patient it is absolutely essential to get the question of compensation settled. My experience of railway cases has convinced me that it is

eminently desirable, so far as the best interests of the patient are concerned, to settle the claim as speedily as possible. I am fully persuaded that in the purely functional cases it is better for the patient to accept a moderate sum in the way of damages at the commencement of the case, than to be subjected to the worry and anxiety attending a prolonged litigation, in the hope (often, when the heavy expenses attending litigation are taken into account, the very uncertain hope) of ultimately gaining a more substantial money compensation.

The Method of Examining Railway Cases.—'Railway cases' differ from the great majority of cases which are met with after ordinary accidents and injuries, in as much as they are usually complicated by the question of money compensation. Since the persons who are injured may claim and be awarded 'damages' in proportion to the severity of the injury, the suffering and loss which it entails and the permanent ill effects which it is likely to leave behind, it seldom, if ever, happens that the symptoms are made light of; on the contrary, there is often unconscious exaggeration and in some cases wilful exaggeration or even deliberate shamming. Cases have actually been known in which persons who were not in the train at all when a collision occurred, have taken to bed, pretended to be ill and claimed damages from the company. Now because of this tendency to exaggeration and deception, railway companies, not unnaturally, regard all cases with suspicion. Medical men are sent to examine the patient and to report to the company. The patient and the patient's doctors, of course, wish to bring additional evidence in their support. The result is, a series of examinations at which several doctors are present, but at which the difficulties of the case are not discussed, and at which, as a rule, little or no interchange of medical opinion takes place. Should the patient and the company fail to come to terms, the case goes into court, and a conflict, or apparent conflict, of medical evidence very often results. These differences of opinion are sometimes very painful; the laity and lawyers, who are unacquainted with the difficulties which some cases of this kind present, distrust the opinion of the doctors and not unfrequently suspect their good faith. It may, however, I think, be unhesitatingly affirmed that the vast majority of medical witnesses who

go into court do so in perfect good faith; their opinion is sometimes biased; occasionally they abandon the neutral ground which unprejudiced scientific witnesses ought to occupy, and assume, or appear to assume, the place of partisans, but they are with rare exceptions witnesses of truth. How comes it then that these differences of opinion and conflicts of medical evidence occur? I propose briefly to mention what appear to be the chief causes, and then, before finally leaving the subject, to detail the manner in which the clinical examination of 'railway cases' should be conducted.¹

1. Differences as to the facts of the case.

The differences of opinion which arise in 'railway cases' are much more frequently differences as to the conclusions to be drawn from the facts, than differences as to the facts themselves. Differences as to the facts do, however, occasionally occur, and they will doubtless continue to occur so long as the present most unsatisfactory method of consultation which is adopted in railway cases continues to be employed. Different individuals are, it must be remembered, both by nature, and by training and experience, very differently qualified in the power of observation. Now in the ordinary consultations of every-day practice the family doctor affords the consultee all the information in his power with regard to the facts and history of the case. Should any difference of opinion arise, either as to the facts themselves or the conclusions to be drawn from those facts, such differences are discussed, the patient is, if necessary, re-examined and an opportunity is afforded for the rectification of any errors which may have arisen on one side or the other. But in 'railway cases' there is, as a rule, little or no interchange of medical opinion. Differences as to facts as well as to the conclusions to be derived from the facts are frequently due to this method of procedure, which ought, as Sir John Erichsen suggests, to be altered. If, for example, the patient's doctors have detected any physical alteration, such as differences in measurement, temperature, electrical reactions and the like, and if these differences have not been detected by the medical men representing the company, the patient's doctor should make known the fact and

¹ The reader is recommended to study the admirable remarks of Sir John Erichsen on this point (*Concussion of the Spine*, Lecture xii.), to which I am indebted for many of the following particulars.

give the railway doctors an opportunity of confirming or rejecting it—in short, the question should then and there be investigated, and, if both sides are satisfied as to the manner in which the observation has been made, they should agree to abide by the result. In this connection it may be well to point out, that conflicting opinions are sometimes due to the fact that delicate clinical tests, such as the electrical testing of muscles and nerves, which require the most scrupulous care of a skilled and experienced observer, are sometimes employed by medical witnesses who are unqualified to use them satisfactorily or to draw correct conclusions from the facts observed. In this way errors both as to fact and opinion may very easily arise. Nor should the patient's doctors, in my opinion, silently acquiesce (as under the present system they almost invariably do) in statements made by the patient which they know to be exaggerated. They ought to see that the company's doctors are correctly and soberly informed as to any facts which have come under their own personal observation. Further, it is highly desirable—and in most cases it can be satisfactorily accomplished without prejudice either to the interests of the patient or the company—to *consult* not merely as to the facts, but as to the conclusions to be derived from the facts. If this were the regular method of procedure, and if, as Sir John Erichsen has suggested, a conjoint report were drawn up and signed by the two opposing sets of doctors, an immense advance in the method of conducting these cases would be made. Sir John Erichsen's suggestions on this point are so important, that I make no apology for quoting them in full. He says—

'The conflict of medical evidence often arises in consequence of a want of proper understanding between the medical men engaged on the opposite sides of the case. As matters are now arranged, there is, as I have already shown, no 'consultation,' in the proper sense of the word, between them. The surgeon of the company examines, it is true, the plaintiff before, and in the presence of his (the plaintiff's) own medical men; but there is no after-discussion of the case, no attempt, as in an ordinary consultation, to reconcile discordant views, and to come to a combined opinion on the case. Neither party knows the exact views of the other on any point, or on the value of any one symptom, until they are heard in court. This is a great evil, and might be corrected by the surgeons on the two sides meeting as ordinary consultants discussing the case together, and, if possible,

drawing up and signing a conjoint report. If such a report could be obtained, it might be handed in for the guidance of the judge and counsel, and the strictly medical part of the case would be much simplified. In fact, it would be disposed of if all parties concerned had substantially agreed before the trial as to the nature, extent, and probable duration of the plaintiff's injuries and their consequences, the tripod on which the medical question always rests. In the event of there being such discrepancy of opinion that an agreement could not be come to on any or all of these points, the judge should appoint at least two surgeons¹ of known character and of recognised skill in the particular class of injury under consideration, to draw up a report upon the plaintiff's past and present condition and future prospects. This report would serve to guide the court in coming to an opinion on the purely surgical part of the case, and afford it that information which men who admittedly know little of a subject on which they are to decide must necessarily be supposed to wish to obtain. The experts or assessors who draw up this report should be appointed by the Court, and not by the litigants. Their position would consequently be an independent one. They could not be accused of unworthy motives. They could not be calumniated, and their evidence would not be disparaged by groundless charges of partisanship.

'The report of such surgical assessors would necessarily be final. It could scarcely be successfully disputed by those medical witnesses from whose conclusions it differed. Hence it would be of paramount importance that none should be selected for such an important post as that of assessor who was not recognised as possessing not only a sound general knowledge of surgery, but such special experience in the diseases resulting from injuries of the cord and brain, as to render his opinion worthy of all consideration in the eyes of his professional brethren. Such a plan would not interfere with the present machinery of the courts. The case would continue to be tried in the ordinary common law courts, before a jury who would decide on all its facts. Their judgment, and that of the court, would be guided in all matters of scientific opinion either by a conjoint surgical report, or, if that cannot be arrived at, by the written statement of competent surgical assessors, who, having had free access to the plaintiff and to the medical reports on both sides, could arrive at a definite and unbiased conclusion as to the nature, extent, and probable duration of his injuries and their consequences. It would, I venture to submit, be in the highest degree advantageous to the medical as well as the legal profession. The great inconvenience of the system of indiscriminately subpoenaing medical practitioners who are but little concerned in the case would be stopped; conflict of medical evidence would no longer occur. Engendered as it is partly by the want of proper understanding

¹ I would venture to substitute for two surgeons—a surgeon and a neurologist. (B.B.)

between the medical witnesses, and greatly encouraged by the want of due scientific knowledge on the part of the court, it would not survive the necessity of both parties either making a conjoint report or submitting their differences of opinion to the arbitrament of skilled surgical assessors selected by the court. And, lastly, the ends of justice would be attained with more certainty than they often are under the present system.' ¹

2. Differences as to the significance of the facts and as to the conclusions to be drawn from the facts.

That differences of this description are of frequent occurrence does not surprise the medical man, however curious and suspicious they may appear to the lawyer and layman. It must be remembered, that in ordinary private practice it is often extremely difficult to determine whether certain symptoms are indicative of functional derangement or organic disease of the nervous system; the most experienced neurologists sometimes have difficulty in forming an opinion on this point. Now, if difficulties in diagnosis and differences of opinion arise in private practice, in other words in cases in which the statements of the patient can generally be relied upon, how much more likely are differences to occur in 'railway cases,' in many of which the symptoms are, in whole or in great part, subjective, and in which there is often much exaggeration and occasionally downright deceit.

Further, it must be remembered that until quite recently the profession was by no means agreed as to the exact significance of many of the symptoms which characterise cases of this description.

Again, under the pressure of counsel, medical witnesses are sometimes induced to give more dogmatic opinions than the facts of the case warrant, or to give opinions on questions with which they are not specially familiar—a physician, for example, may under the pressure of counsel give an opinion on a question which is surgical rather than medical, and *vice versa*.²

¹ *Concussion of the Spine*, page 324, *et seq.*

² This statement is of course general. There are doubtless some physicians who are able to give good opinions on surgical questions, just as there are certainly some surgeons who are well qualified to give opinions on medical subjects. But, speaking generally, the same rule, which in private sends medical cases to the physician and surgical cases to the surgeon, should be followed in railway practice. In ordinary private practice difficult nervous cases are referred to the neurologist. The same procedure should be followed in railway cases. It is certain that if this were done there would be far less conflict of opinion in court.

Then again, in many cases the physician or surgeon who is called in as an expert—and the statement applies perhaps with greater force to the experts sent by the company than those called in by the patient—has only one opportunity of seeing the patient, and as the result of that one examination he is expected to pronounce a decided opinion upon all the facts of the case. Doubtless in private and also in railway practice, one thorough examination is usually quite sufficient to enable a consultant to form an opinion; but cases do every now and again occur in which the most competent practitioner withholds his opinion until he has had further opportunities of studying and watching the case; and there are no cases in which such opportunity of further observation is more necessary than in some of the cases in which a differential diagnosis of functional or organic disease of the nervous system has to be made.

Speaking generally, **the method of examination which is to be followed in railway cases** is the same as that which we are in the habit of employing in ordinary private or hospital practice. There are, however, certain differences in detail which it is necessary to remember and observe.

The examination must be very full and elaborate, and very careful and accurate notes must be taken of each fact as it is elicited. This necessitates a very prolonged examination; and I may observe in passing, that the series of lengthy examinations which railway patients have to go through, cannot fail, in some instances, to exert a prejudicial effect upon their condition.

An exact description must be obtained from the patient—and it often requires a good deal of skilful cross-examination to elicit all the particulars—of:—(1) the accident (the exact manner in which he was hurt, the sensations which he experienced at the time, the effects which immediately followed, the manner in which he was able to conduct himself immediately afterwards, etc.); (2) the symptoms which he experienced, and the treatment which was adopted during the acute stage of the illness, i.e. during the first week or two after the accident; (3) the mode of development of the after-symptoms, and the whole subsequent progress of the case up to the date of the examination.

The patient's private medical attendant should next detail the facts which he himself observed when the patient came

under his care, the course of treatment which was adopted and the subsequent medical history of the case.

It is also of the greatest importance to ascertain, both from the patient himself, his friends and his medical adviser, the exact condition of health before the accident; for, as has been previously pointed out, patients are very apt to attribute to an accident or injury diseased conditions which were previously present, but which had not been previously observed.

It is especially important to endeavour to arrive at a just estimate of the patient's ordinary nerve tone and nerve balance. The nerve tone and nerve balance of his near relatives should also be carefully inquired into, for, as we all know, a tendency to nervous disturbance is much more strongly developed in some families than in others.

Having completed the inquiry into the history of the case, the condition of the patient at the time of the examination must be carefully and fully investigated. Care must be taken to describe the facts in the simplest language and to avoid any terms which imply theories as to the facts. A patient, for example, may state that he is unable to move his legs. Such a condition may be due either to the fact that he *will* not (does not choose to) put the muscles into action, or to paralysis. The bare fact that the patient says he cannot move the limbs should be noted; whether the inability depends upon paralysis, and whether that paralysis depends upon organic disease or mere functional disturbance, are conclusions which, in many cases, cannot be correctly determined until all the facts and circumstances of the case have been taken into careful consideration.

In forming a judgment as to the exact value of the symptoms, it must be remembered, that railway cases are usually complicated by the question of money compensation, that in many cases there is unconscious exaggeration, and that in some cases there is wilful exaggeration and deception. Subjective symptoms *per se* should always be viewed with suspicion and should be valued according to the character and reliability of the patient, his general behaviour, the amount of damages which he is claiming and the whole collateral (non-medical) facts of the case. On the other hand, more importance (relatively to the subjective symptoms) should be attached to objective signs of disease than in private or hospital practice. Care must, how-

ever, be taken not to give too much weight to slight objective alterations (such, for example, as slight differences in temperature on the two sides of the body), more especially when other positive signs of organic disease are wanting. As I have previously insisted upon, no importance should be attached to slight objective differences obtained by instrumental means, such as electricity, unless they have been elicited by an observer whose capabilities of observation are beyond suspicion. Any anomalous symptoms or complaints which seem to be out of harmony with the other facts of the case should be allowed due weight, and should be prominently noted in the report. Above all things, the medical expert should endeavour to forget that he is called by this side or by that; after having carefully and conscientiously elicited *all* the facts, he should endeavour to form an unprejudiced judgment and to take into account the probabilities both for and against the patient. He is sworn, be it remembered, when in court, to speak the truth, *the whole* truth, and nothing but the truth.

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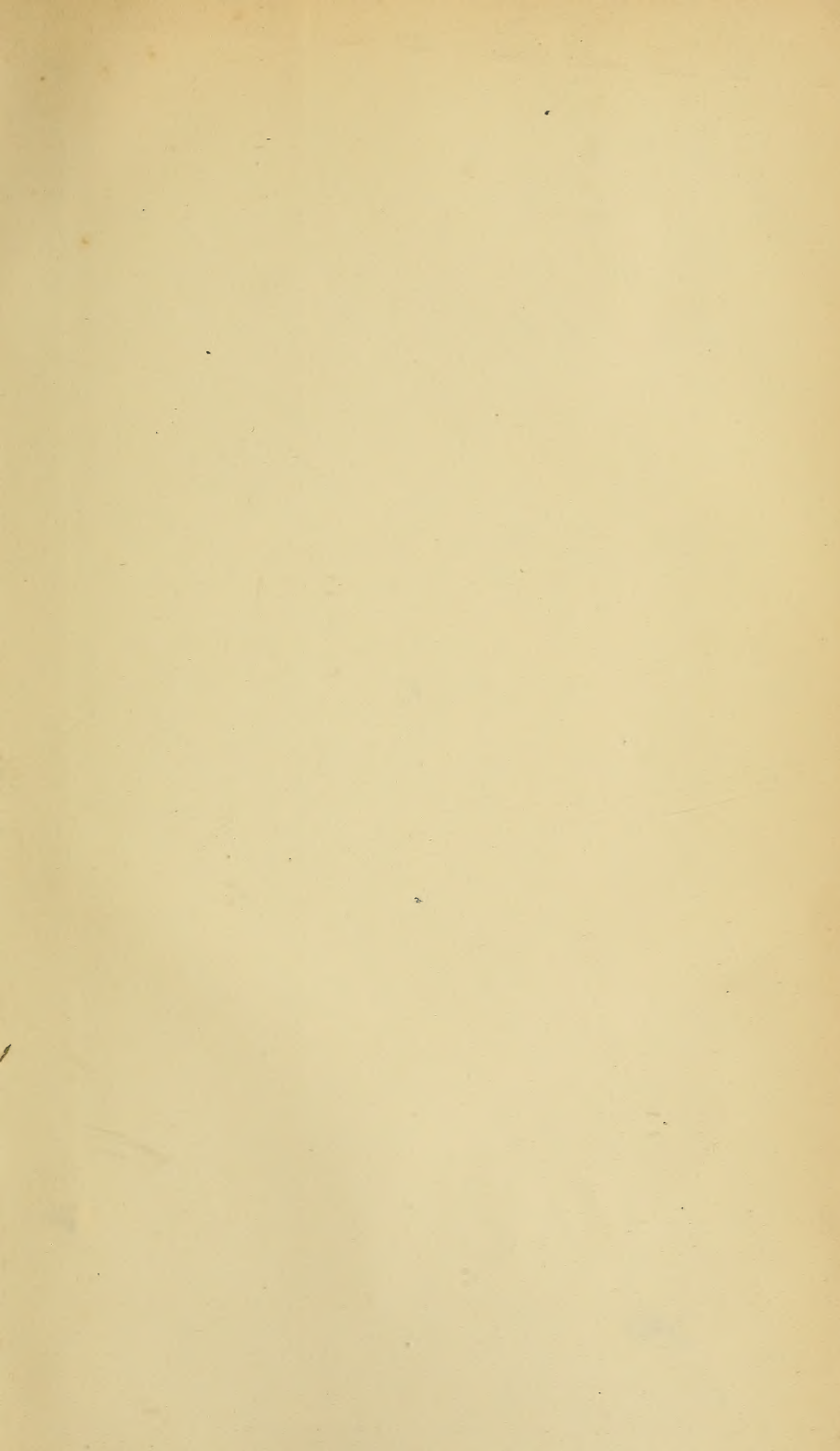
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